FAKULTNÍ NEMOCNICE OLOMOUC

icní arteriální hypertenze –

Martin Hutyra

pertension

Treatment of Pulmonary
ardiology (ESC) and the

iatric and Congenital
r Heart and Lung

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Cardiovascular Institute, Guangdo
Guangzhou, China. [5]Division of P
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University of Michigan, Ann Arbor
Hôpital Bicêtre (AP-HP), Le Kreml

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ESC European Society of Cardiology

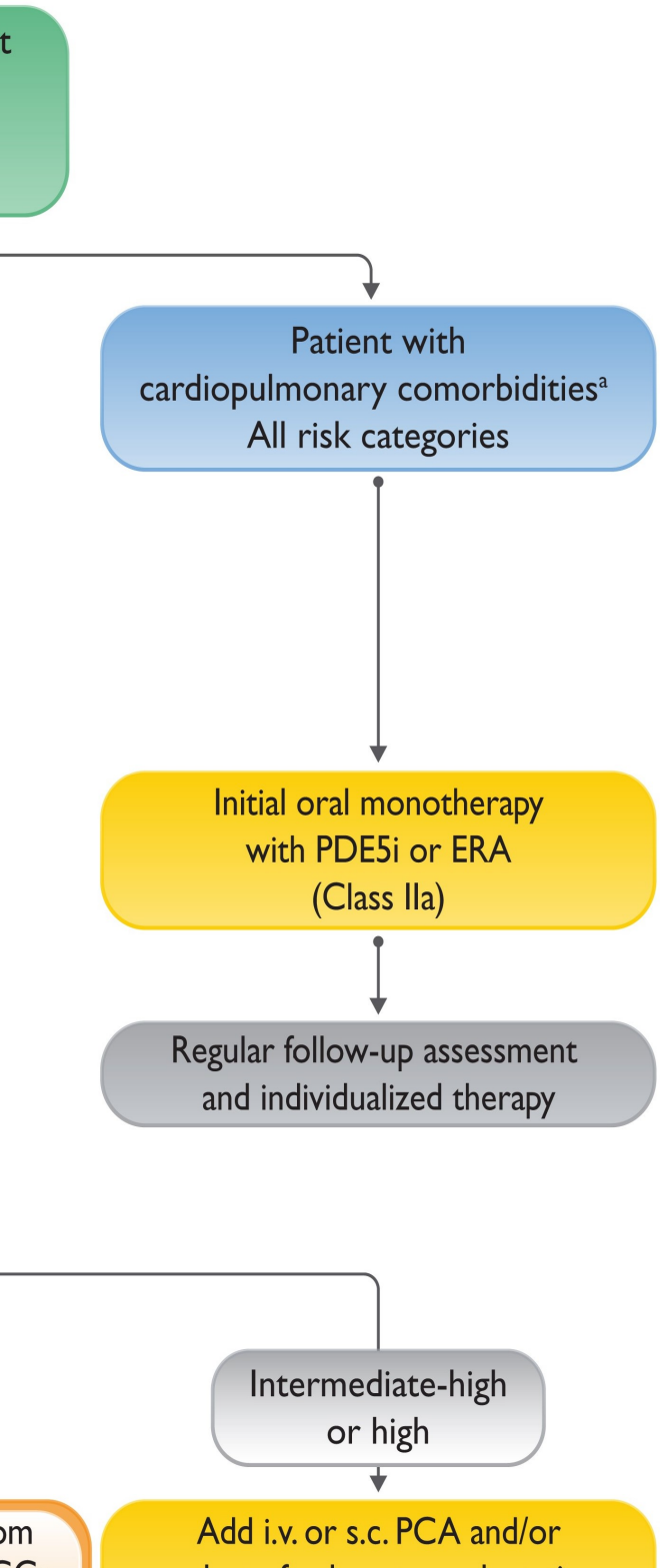
European Heart Journal (2022) **00**, 1–114
https://doi.org/10.1093/eurheartj/ehac237

ESC/ERS GUIDELINES

22 ESC/ERS Guidelines for the diagnosis and eatment of pulmonary hypertension

veloped by the task force for the diagnosis and treatment of monary hypertension of the European Society of Cardiology

No	Slow	Rapid
No	Occasional syncope[a]	Repeated syncope[b]
I, II	III	IV
>440 m	165–440 m	<165 m
Peak VO_2 >15 mL/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 mL/min/kg (35–65% pred.) VE/VCO_2 slope 36–44	Peak VO_2 <11 mL/min/kg (<35% pred.) VE/VCO_2 slope >44
BNP <50 ng/L NT-proBNP <300 ng/L	BNP 50–800 ng/L NT-proBNP 300–1100 ng/L	BNP >800 ng/L NT-proBNP >1100 ng/L
RA area <18 cm^2 TAPSE/sPAP >0.32 mm/mmHg No pericardial effusion	RA area 18–26 cm^2 TAPSE/sPAP 0.19–0.32 mm/mmHg Minimal pericardial effusion	RA area >26 cm^2 TAPSE/sPAP <0.19 mm/mmH Moderate or large pericardial effusion
RVEF >54% SVI >40 mL/m^2 RVESVI <42 mL/m^2	RVEF 37–54% SVI 26–40 mL/m^2 RVESVI 42–54 mL/m^2	RVEF <37% SVI <26 mL/m^2 RVESVI >54 mL/m^2
RAP <8 mmHg	RAP 8–14 mmHg	RAP >14 mmHg



(ESC) and the European

Endorsed by the Interna Transplantation (ISHLT) on rare respiratory disea

Recommendations
During follow-up
In patients with IPAH/HPAH/DPAH who present at in while receiving ERA/PDE5i therapy, **addition of selex**
In patients with IPAH/HPAH/DPAH who present at in of death while receiving ERA/PDE5i therapy, **additio** analogues and **referral for lung transplantation** eval
In patients with IPAH/HPAH/DPAH who present at in while receiving ERA/PDE5i therapy, **switching from** considered

Determinants of prognosis	Low risk	Intermedia risk
Points assigned	1	2
WHO-FC	I or II	-
6MWD, m	>440	320–44
BNP or	<50	50–19

, B6, **NT-proBNP** 697,5 ng/L

KOMORA bez dilatace a hypertrofie, s normální syst

EF LK 55-60%. Diastolická funkce: porucha relaxace

SA 15,8 cm2, FAC PK 18%, TAPSE 16 mm, Vt 13 cm

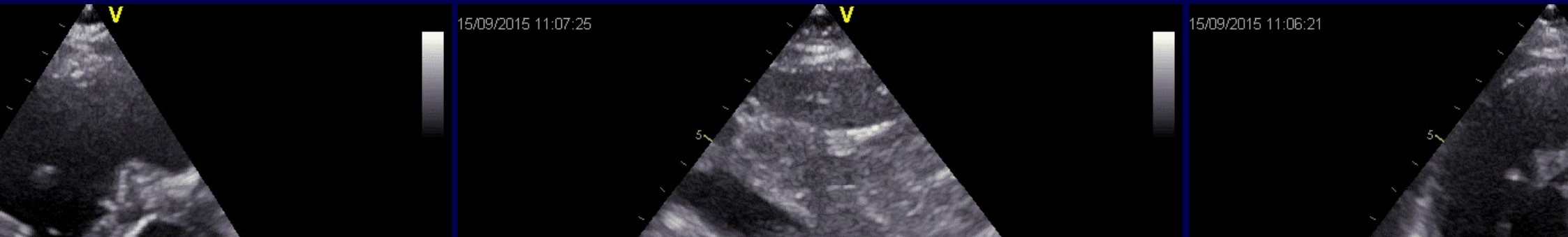
adovaný PASP 90 mmHg, MPAP 55 mmHg, **TAPSE/**

mmHg, svO2 58,6%, PK: 82/10 mmHg, AP: 84/41/5

CO: 3,43 l/min., CI: 1,81 l/min./m2, **SVI 26 ml/m2**

egativní

ez ventil. poruchy, TLCO 67% NH

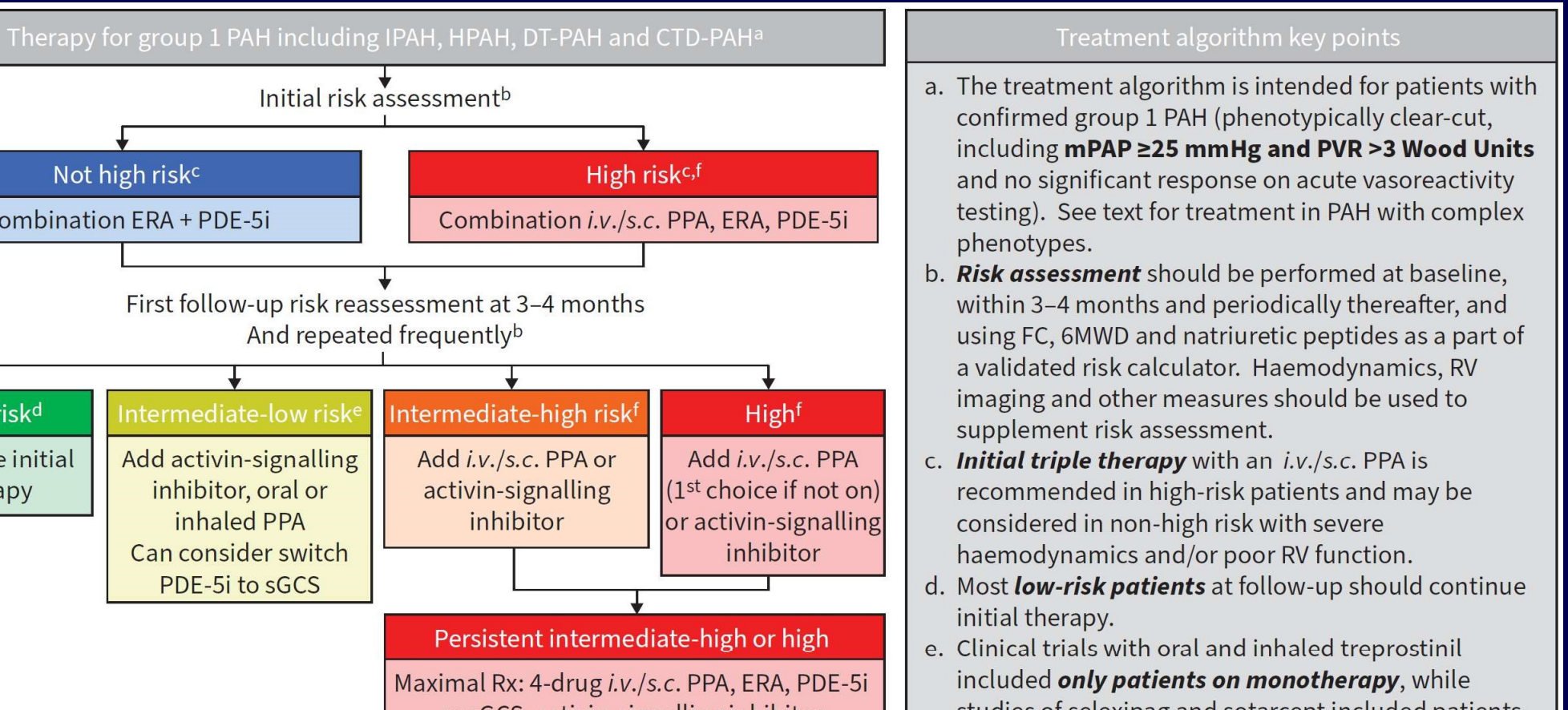


terapie ERA + iPDE5

jící selexipag + ERA + iPDE5

rnující prostacyklin s.c./i.v. + ERA + iPDE5

s časnou eskalací dle efektu a rizikové stratifikace



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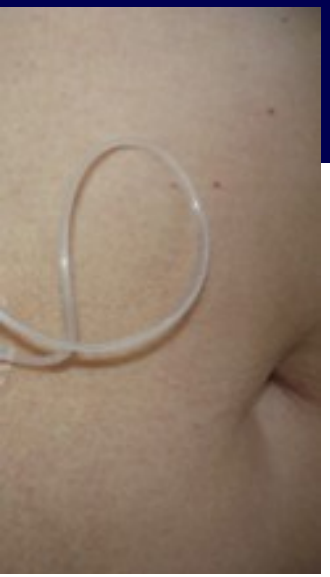
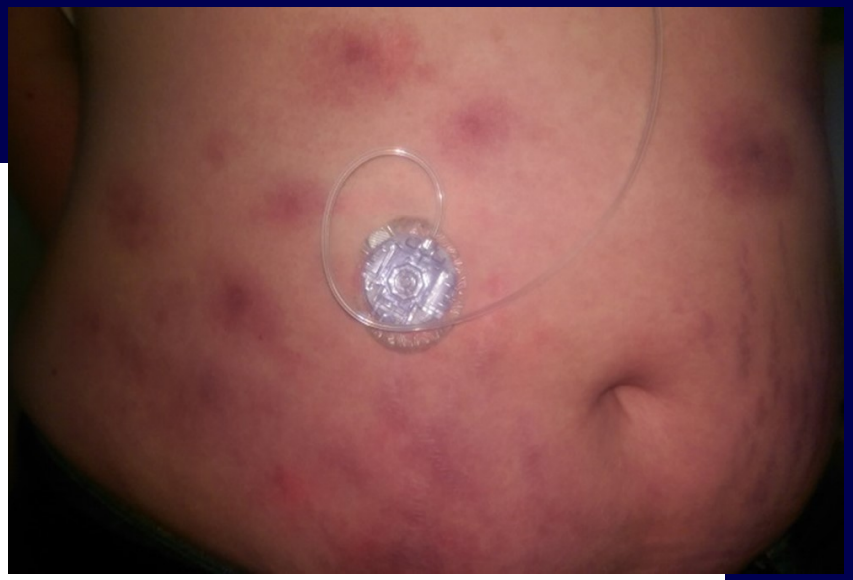
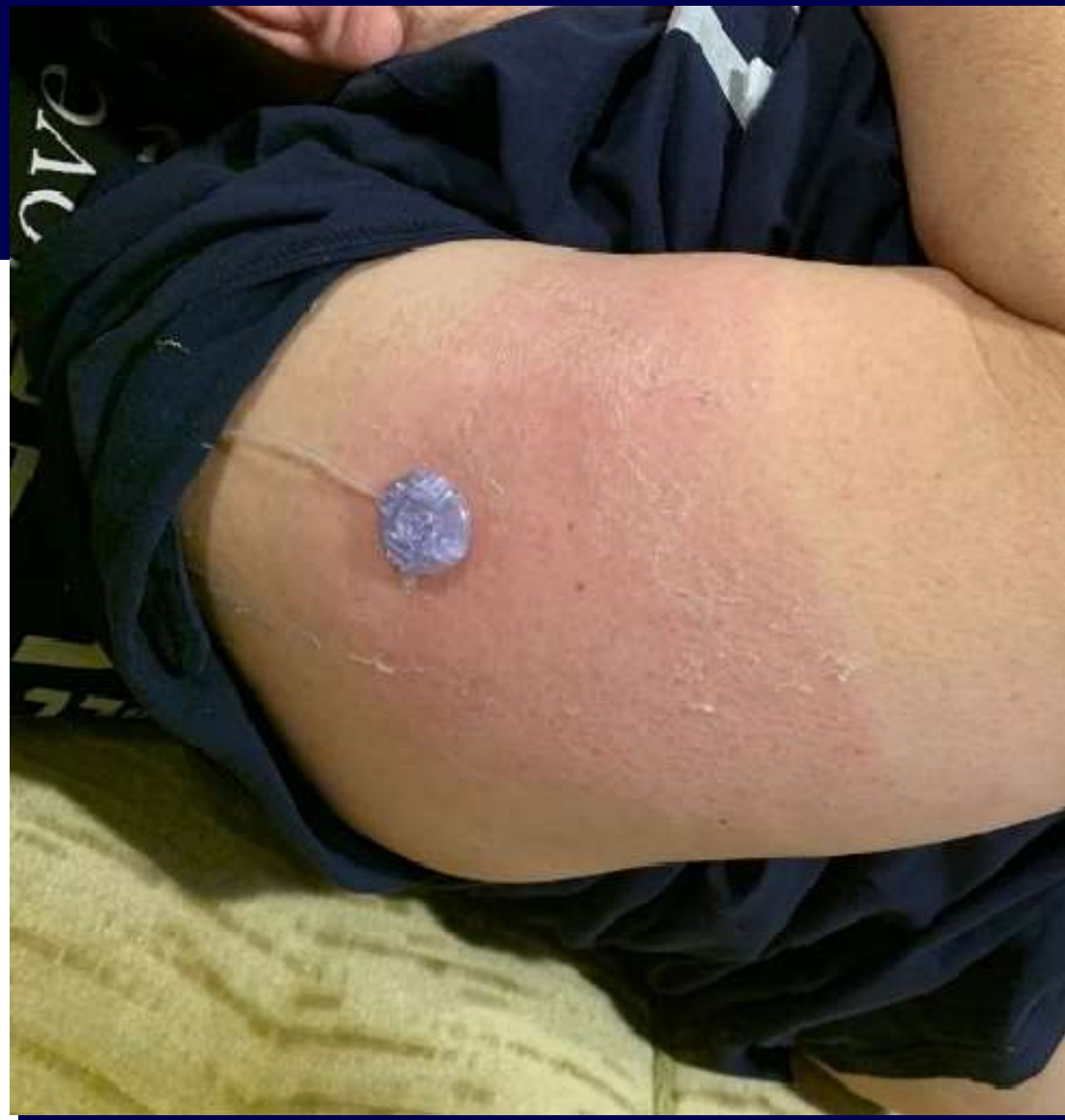




NT-proBNP [0;125]

avky prostacyklinu sítí s přiměřenou lokální a
optimálního místa aplikace + ERA + iPDE5
a převod na p.o. trojkombinaci zahrnující selexipag
jící prostacyklin i.v. v cílové dávce + ERA + iPDE5



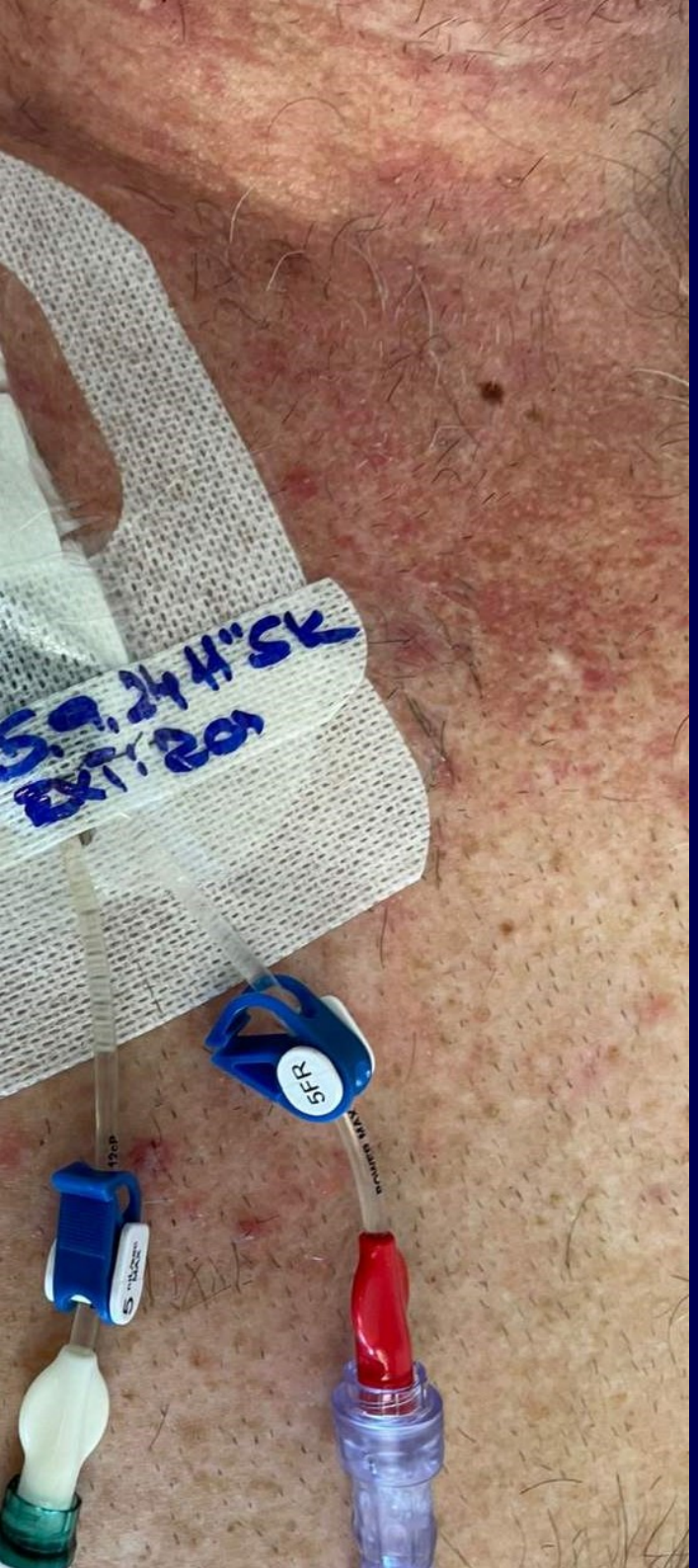
NT-proBNP [0;125]

Horní mez

pumpy s i.v. aplikací treprostinilu

a převod na p.o. trojkombinaci zahrnující selexipag

rnující prostacyklin i.v. v cílové dávce + ERA +



Plicní hyper[redacted]

Ahoj zlatíčka. Prosí
přepichu máte bole
den a už tu bolest n
ustupovala tak se s
Nemůžu sedět,choc

To se mi líbí

3

Zobrazeno 51 uživateli

Nejlepší komentáře

[redacted]

Už rok si da
dobu jsem
dny a rozho
přepichu do
hodně vyde

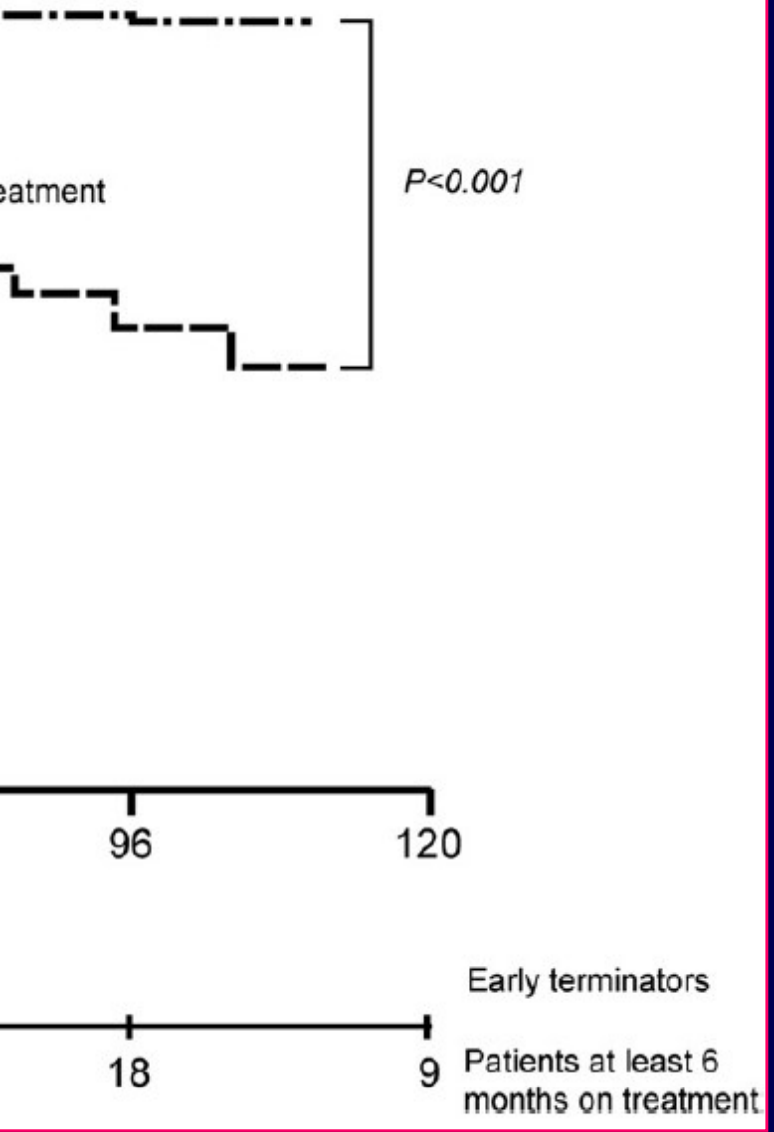
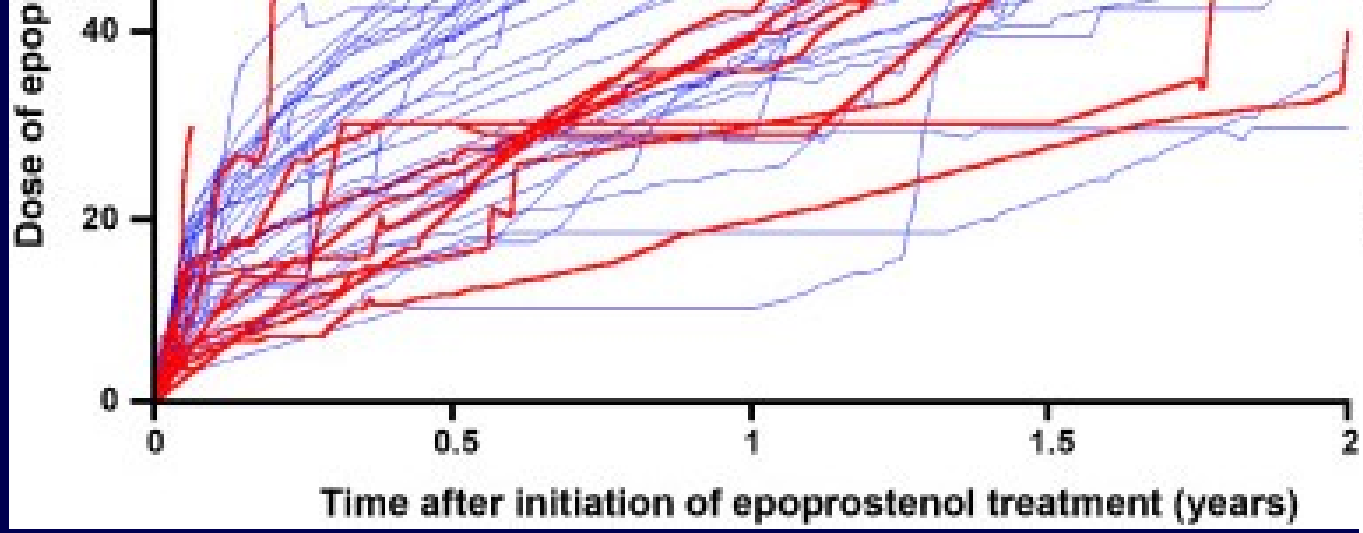
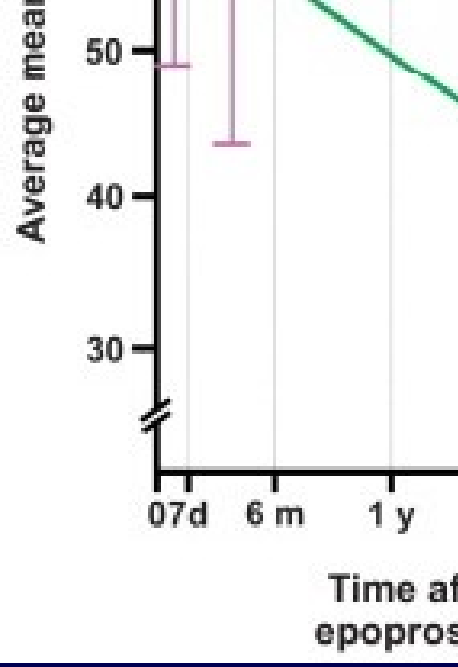


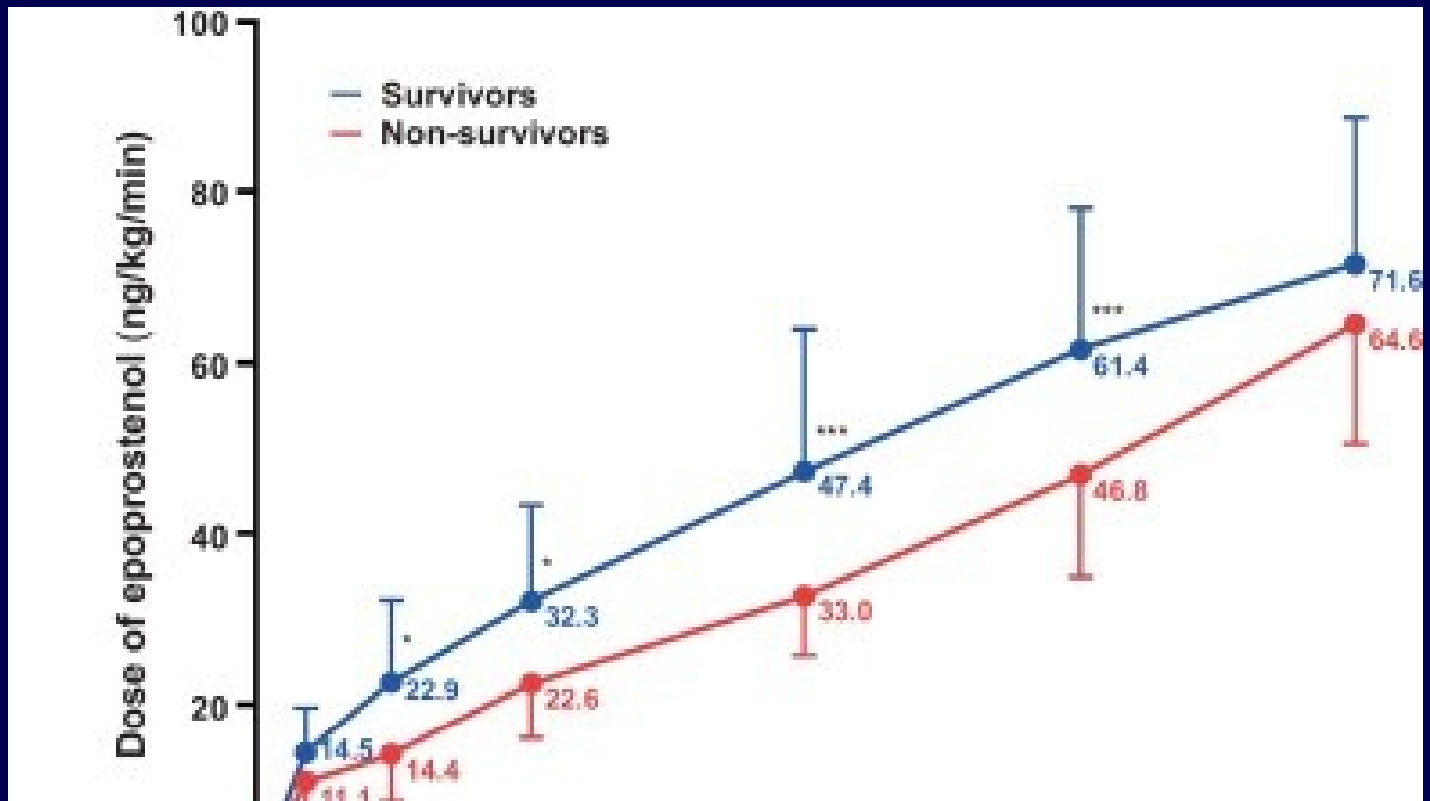
Table 6 Multivariate Predictors of Survival

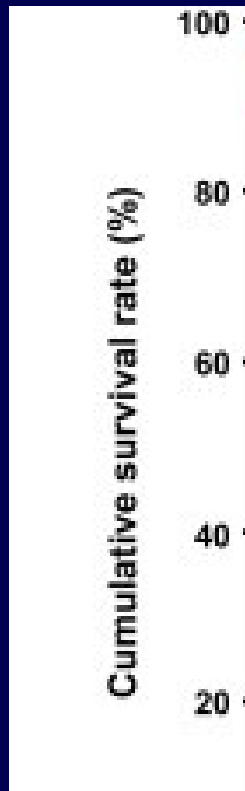
Prognostic factor	Hazard ratio for death (95% CI)	*p*-value
Patient-related factors (based on available data for 811 patients)		
Age, ↑ 10 y	1.01 (0.89–1.15)	NS
BMI, ↑ 10 kg/m^2	0.52 (0.38–0.72)	<0.001
Albumin, ↑ 0.5 g/dl	0.70 (0.58–0.83)	<0.001
Sodium, ↑ 5 mmol/liter	0.90 (0.70–1.15)	NS
Total bilirubin, ↑ 0.1 mg/dl	1.02 (1.00–1.03)	0.033
Creatinine, ↑ 0.5 mg/dl	1.73 (1.31–2.28)	<0.001
On-treatment factors at Week 12		
Treprostinil dose[a] ↑ 10-ng/kg/min increments	0.64 (0.45–0.89)	0.009
6MWD[b] ↑ 20-m increments	0.86 (0.78–0.95)	0.004
PVRI[c] ↓ 10 mm Hg/liter/min/m^2	0.73 (0.44–1.21)	NS

ors	*p* value
	0.301
	1.000
	0.849
	0.133
	0.01
	0.010
	0.006
	0.272
	0.850
	0.066
6	0.107
	0.279
9	0.080
	1.000
	1.000







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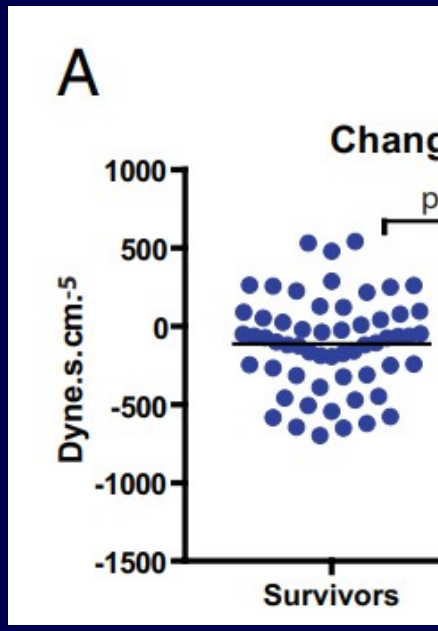
A
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-500
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-1500
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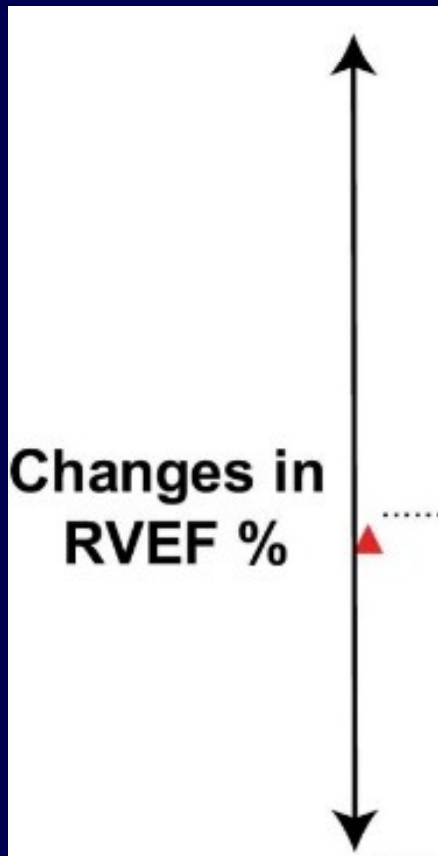
Survivors


Changes in
RVEF %

DĚKUJEME ZA POZORNOST

FAKULTNÍ NEMOCNICE OLOMOUC

UIDELINES PRES WSPH 2024 S VÝHLEDEM D

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KOMPLEXNÍ
KARDIO
VASKULÁRNÍ

European
Reference
Network
for rare or low prevalenc
complex diseases

Network

on of isolated
of
ry arteries

old patient
utopsy: pulmonary
r sclerosis,
e RV hypertrophy

Department of Pathology, University of Liverpool

ex (2-amino-5-phenyl-2-oxazoline) is an appetite-suppressing drug which was availab
and from November 1965 to October 1968. In 1967 a sudden 20-fold increase i
ce of primary pulmonary hypertension was observed in a Swiss medical clinic. It
that a considerable number of these patients had taken aminorex to reduce weig
increase in the incidence of primary pulmonary hypertension was encountered in
n Switzerland, and also in Austria and Germany, where aminorex was available
d incidence of the disease has not been reported in countries where this drug wa
e. A decline in the incidence of primary pulmonary hypertension has occurred in Sw
ce the withdrawal of aminorex. We have administered a high oral dose of aminor
up to 43 weeks and to dogs for 20 weeks. A detailed quantitative pathological exa
the heart and pulmonary vasculature in these animals has failed to reveal any eviden
nsive pulmonary vascular disease. Although there is statistical evidence linking ami
lmonary hypertension, there is no proof that aminorex causes hypertensive pulm
disease in man. It is nevertheless important to enquire into the diet and history of
n in any patient with unexplained pulmonary hypertension.

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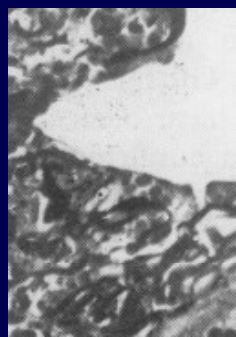

t on a WHO meeting
va, 15–17 October 1973

Edited by

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and

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orld Health Organization,
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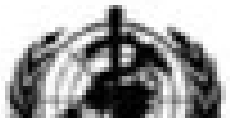

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Dr J. A. Will, Associate Professor, Department of
cultural and Life Sciences, University of Wisco

Dr P. Wirz, Department of Internal Medicine, Med

CONTENTS

Etiological classification

The clinician identifies abnormally high p
by a combination of bedside and catheterization
to diagnose a particular cause (*pulmonary hy*
Most conditions known to cause chronic cor
belong to this category. It includes chronic pu
or pulmonary veno-occlusive disease if identifie
due to heart diseases such as left-to-right shunt
are excluded by definition from causes of co
also important in this category. In other case
able to identify the etiology (*pulmonary hype*
traditionally known as "primary pulmonary
assigned to this category only when all possib
excluded. In some cases the etiological agent m
hypertension of doubtful cause).

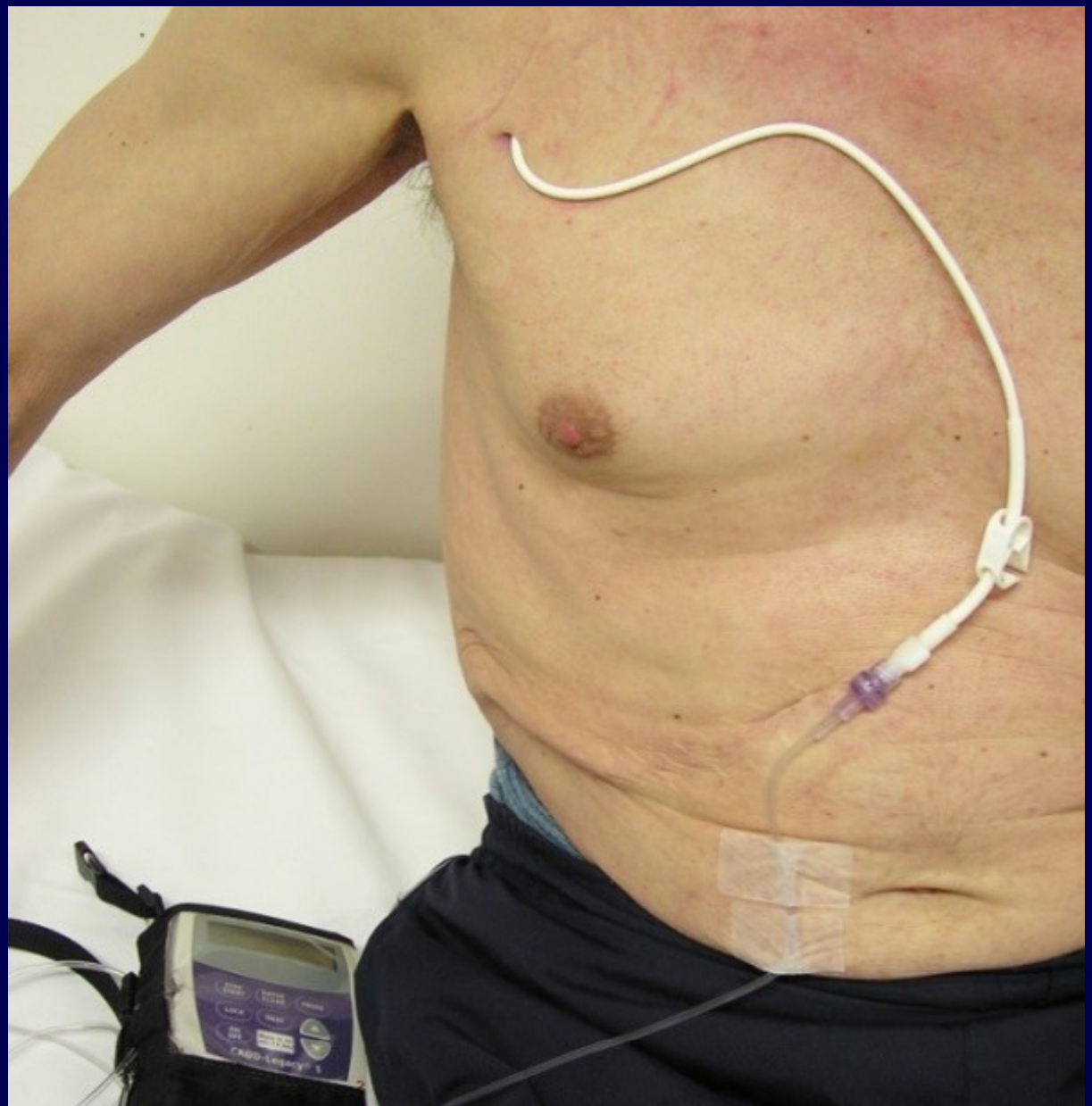
Morphological classification

Various morphological changes are obser
upon the cause (see page 14). When the cause
histopathological patterns may be identified:

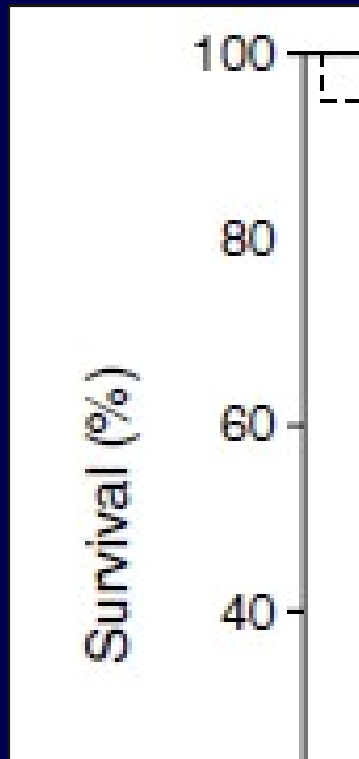
(*a*) pulmonary vascular disease characteri
fibrosis, necrotizing arteritis, and plexiform le

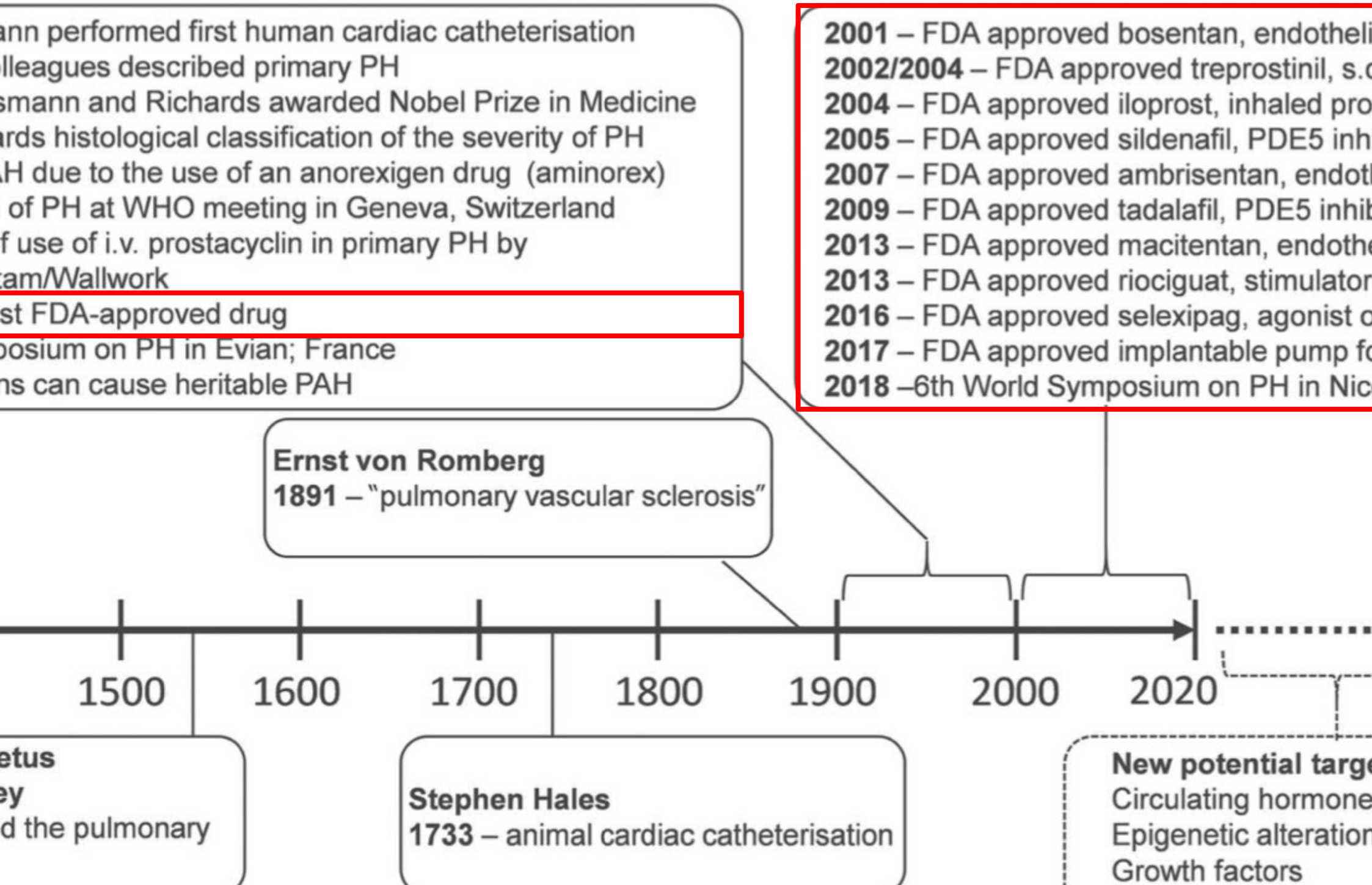
RRY F. URETSKY, M.D., LINDA M. CLAYTON, PHARM.D., MARIA M. JÖBSIS,
MER D. BLACKBURN, JR., B.A., DENISE SHORTINO, M.S., JAMES W. CROW,
FOR THE PRIMARY PULMONARY HYPERTENSION STUDY GROUP*

	CONVENTIONAL THERAPY (N = 40)
OSTENOL = 41)	
0±3	40±2
(24)	12 (30)
(76)	28 (70)
(76)	29 (73)
(24)	11 (28)
2±8	25±6
(66)	24 (60)
1±2	59±2
3±1	12±1
0±2	89±2
0±1	10±1
0±0.1	2.1±0.2
9±2	86±2



VARIABLE
Pulmonary vascu sistance (mm liter/min)




ann performed first human cardiac catheterisation
lleagues described primary PH
smann and Richards awarded Nobel Prize in Medicine
ards histological classification of the severity of PH
AH due to the use of an anorexigen drug (aminorex)
of PH at WHO meeting in Geneva, Switzerland
f use of i.v. prostacyclin in primary PH by
tam/Wallwork
st FDA-approved drug
posium on PH in Evian; France
ns can cause heritable PAH
2001 – FDA approved bosentan, endotheli
2002/2004 – FDA approved treprostinil, s.c
2004 – FDA approved iloprost, inhaled pro
2005 – FDA approved sildenafil, PDE5 inh
2007 – FDA approved ambrisentan, endot
2009 – FDA approved tadalafil, PDE5 inhib
2013 – FDA approved macitentan, endothe
2013 – FDA approved riociguat, stimulator
2016 – FDA approved selexipag, agonist o
2017 – FDA approved implantable pump fo
2018 –6th World Symposium on PH in Nic
Ernst von Romberg
1891 – "pulmonary vascular sclerosis"
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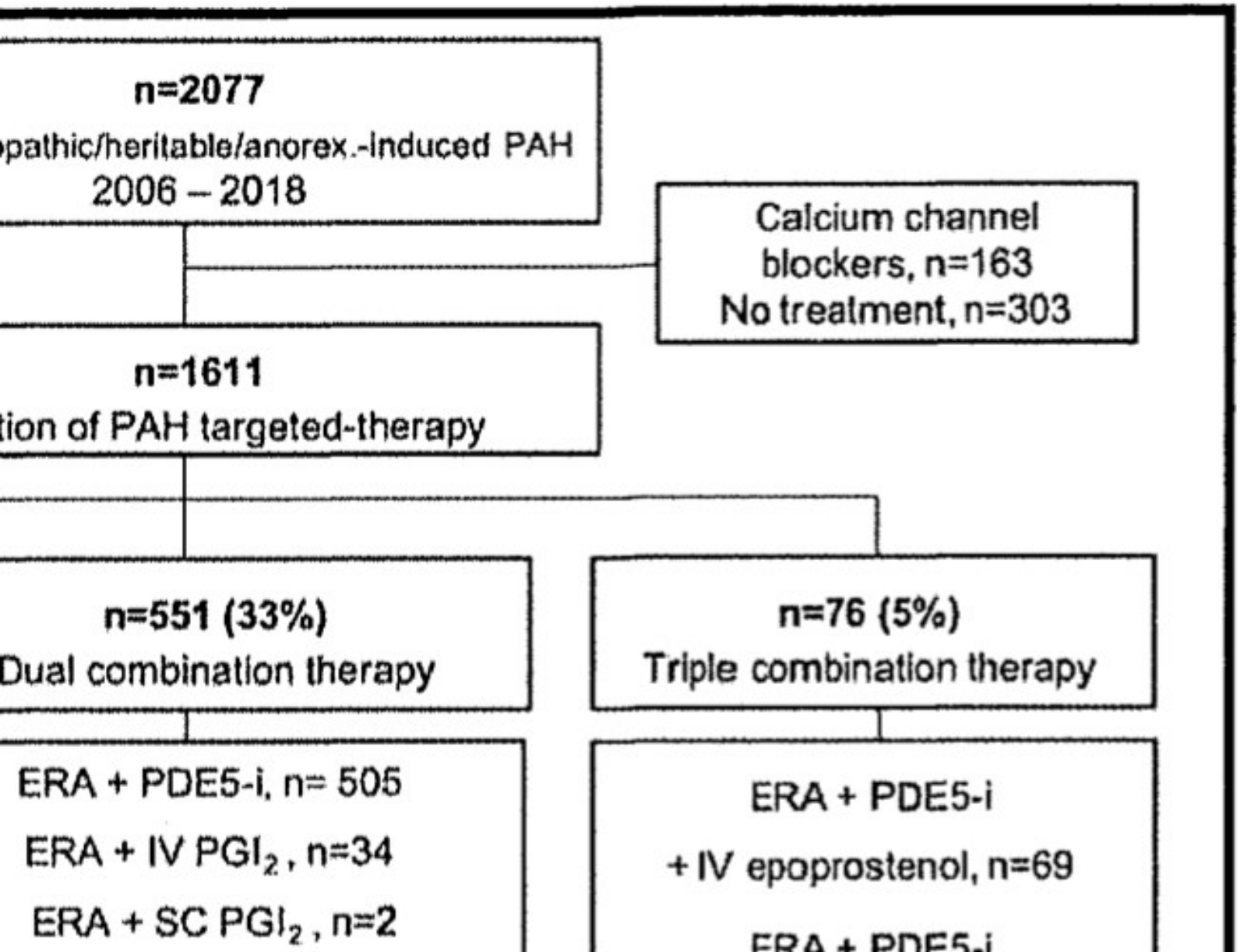
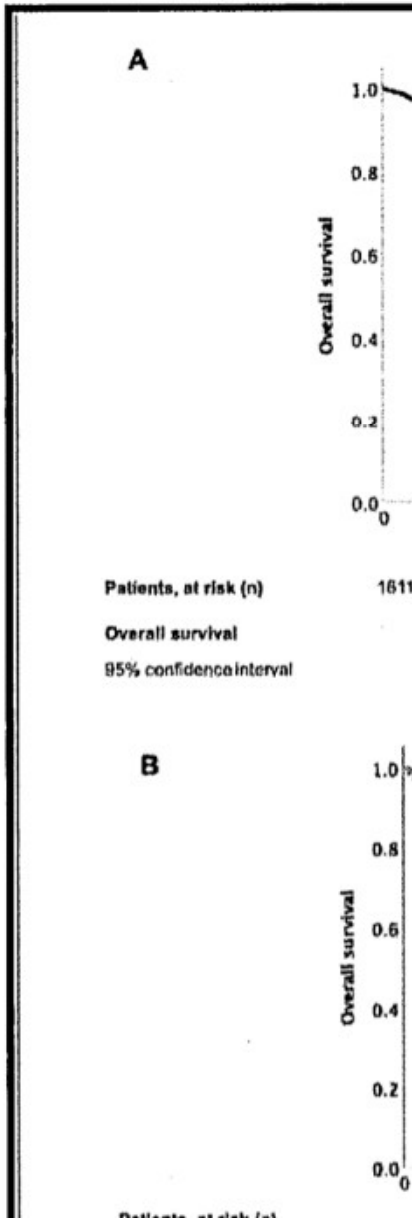
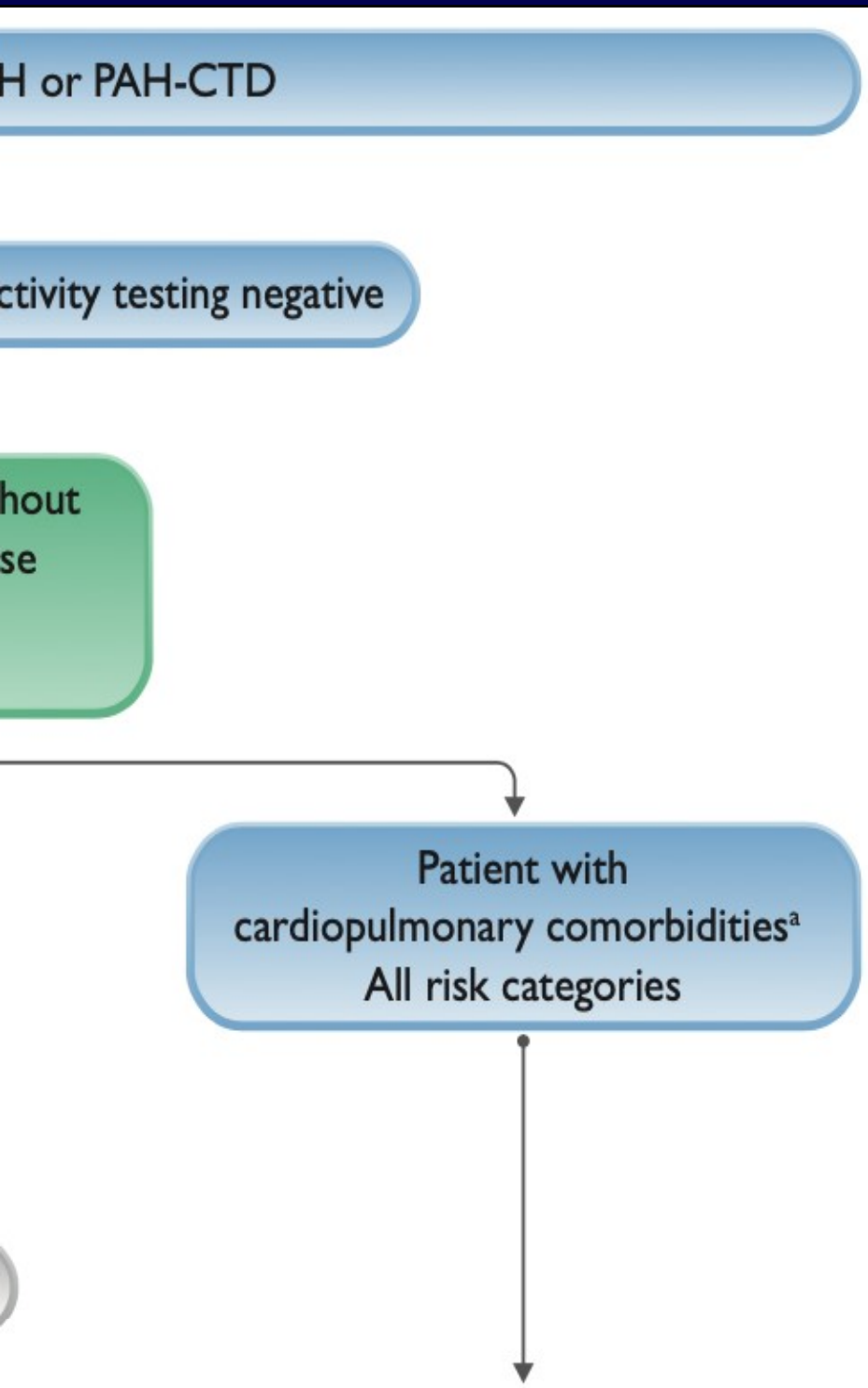
d the pulmonary
Stephen Hales
1733 – animal cardiac catheterisation
New potential targe
Circulating hormone
Epigenetic alteration
Growth factors

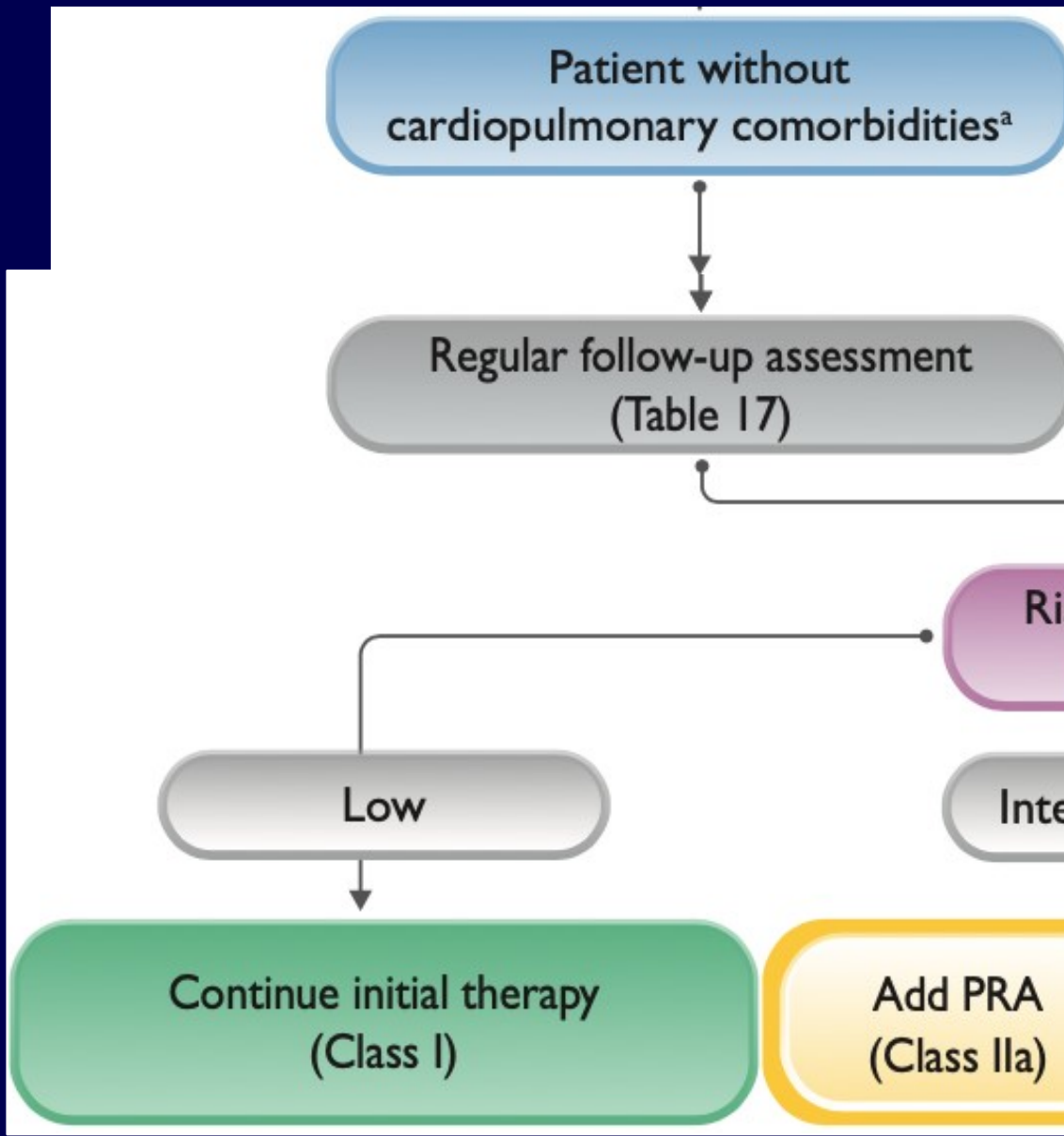
006-2018

otherapy, 551 dual combination therapy, 76 triple cpm

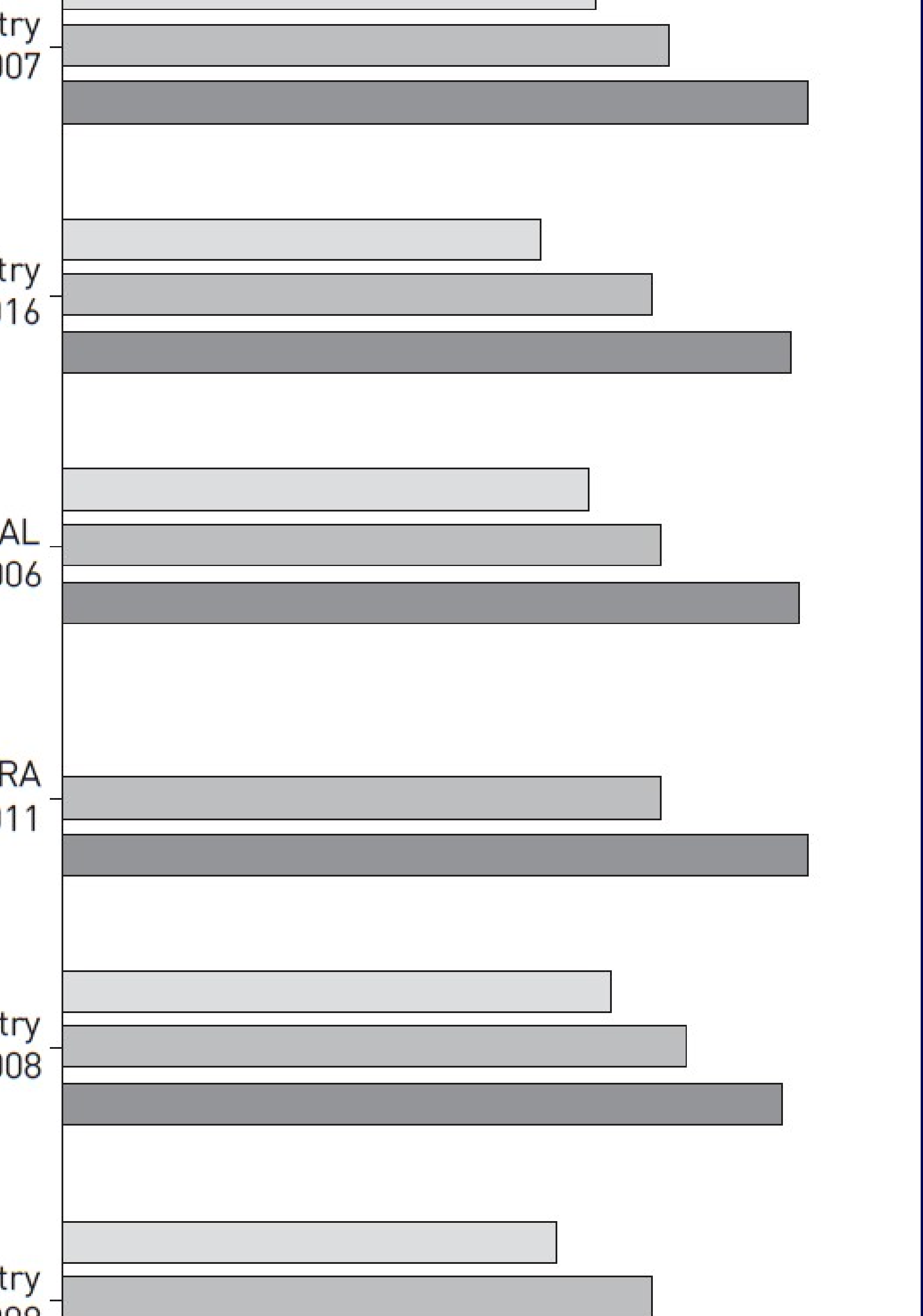








Determinants of prognosis	Low risk	Intermed
Points assigned	1	


try
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rs,187 patients diagnosed between 1981 and 19
emale 1:1.7, FU through 1988 (106 died)
ptoms: dyspnea (60 %), fatigue (19 %), syncop

MEDICAL AN

Forty-five pe
rent cigarette s
petite suppressa
male patients ha
There were 2.3
try. None of th
cally from thos
were 12 cases
(disease affecti
and 5 in wome

l 2,967 US adult patients with PAH from March 2
toms were experienced for >2 years before PAH

racteristics

	>2 y (n = 526)	*P* Value[a]
7)		
8)	160 (30.4)	< .001
1)	109 (20.7)	.049
0)	118 (22.4)	.046
0)	93 (17.7)	.003
2)	46 (8.7)	Reference
6)	105 (20.0)	.53[b]
4)	421 (80.0)	

Comor

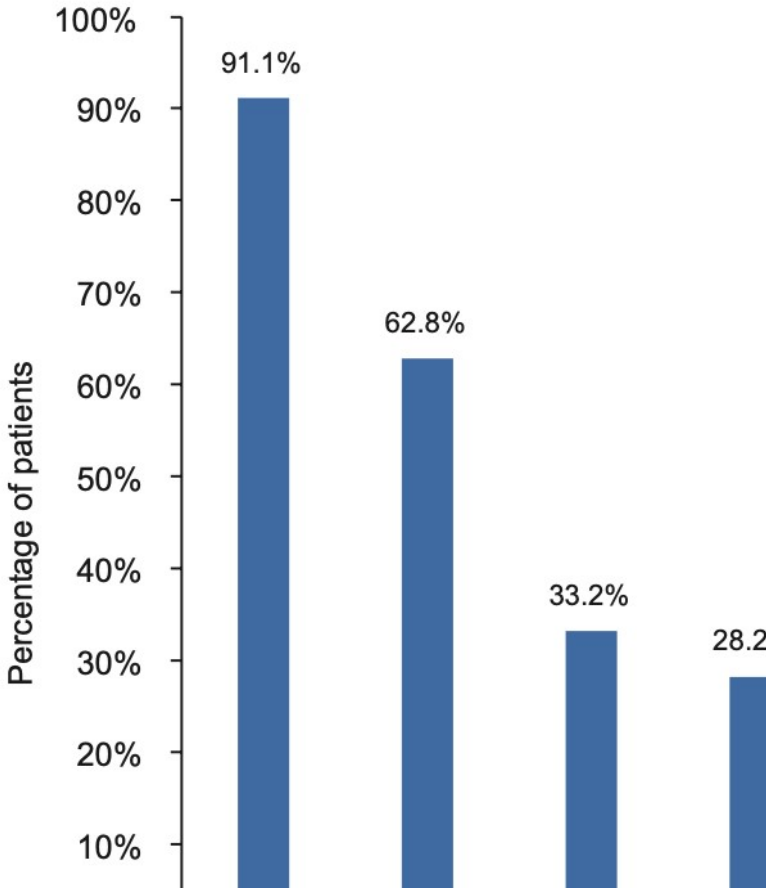
Characteristic
Comorbid conditions at diagnosis of PAH
History of obstructive airways disease[b]
History of thromboembolic disease
Sleep apnea
Obesity (BMI ≥ 30 kg/m²)

ey conducted in five European countries (EU5), 572 patients

ns to diagnosis

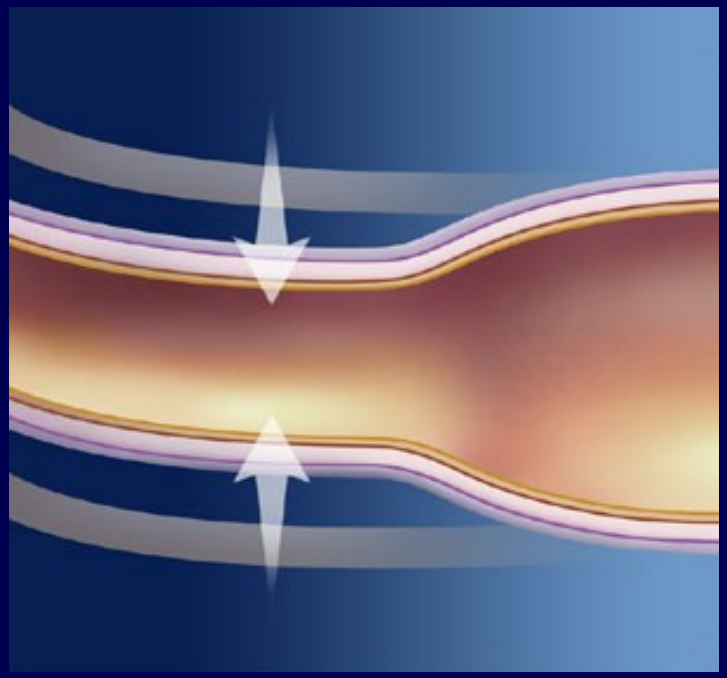
S

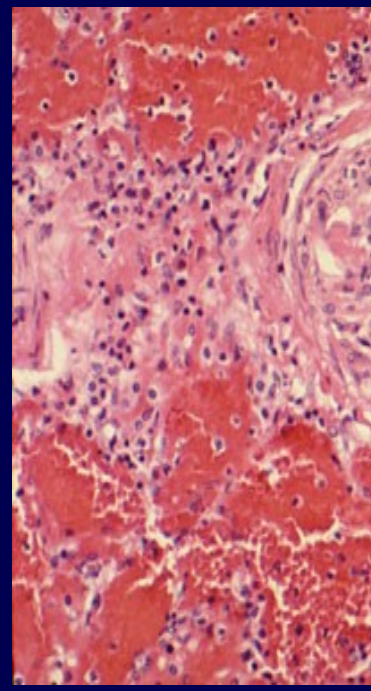
	Country		
	EU5	USA	Japan
	n = 305 7.8 (0.2, 120.0)	*n* = 153 15.3 (0.5, 120.0)	*n* = 72 5.1 (0.2, 60.0)
	n = 317 7.8 (0.2, 155.9)	*n* = 154 8.6 (0.2, 60.0)	*n* = 74 5.5 (0.2, 72.0)
	n = 290 15.1	n = 152 24.0	*n* = 70 9.9

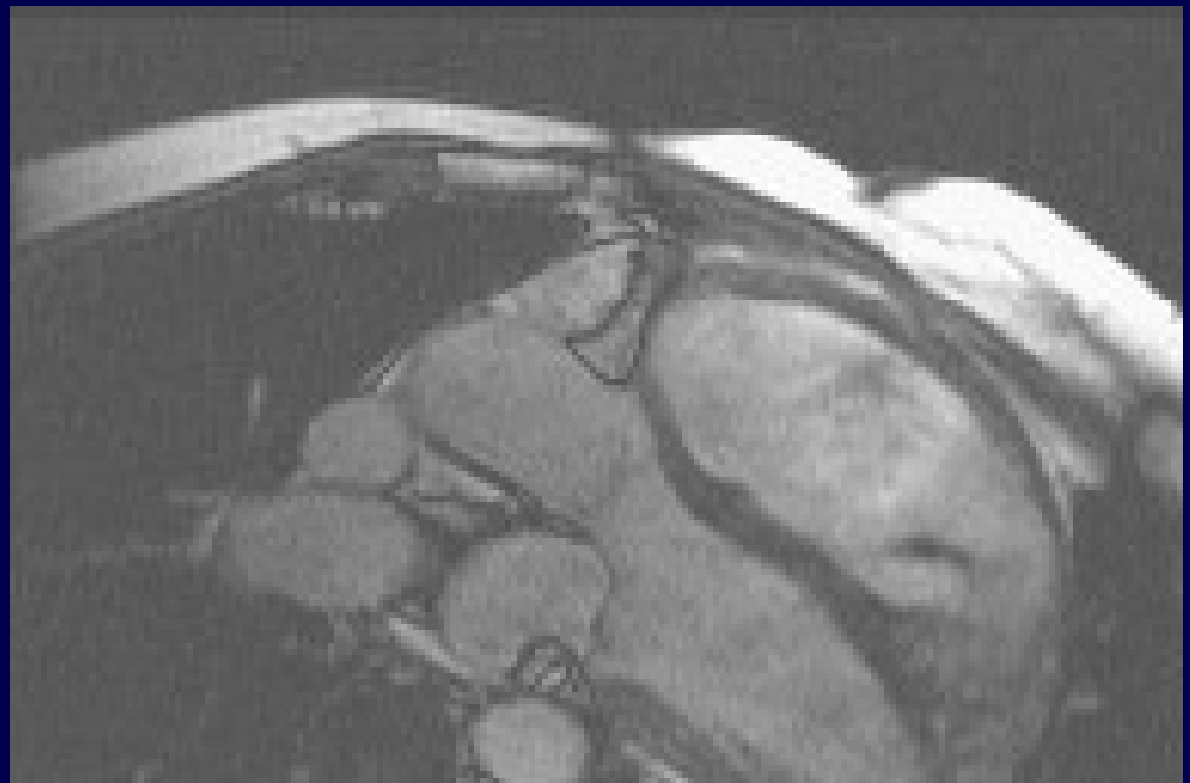


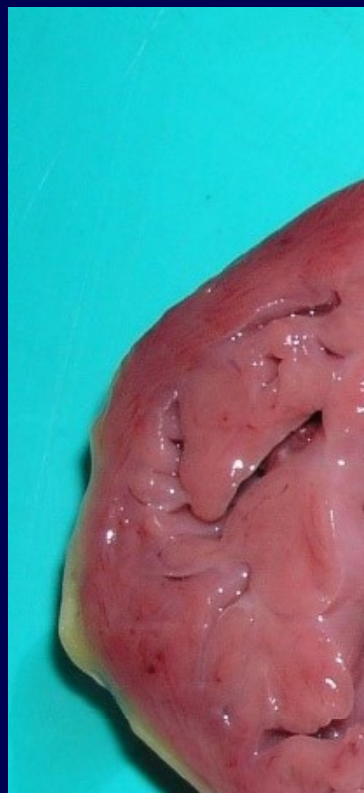
halis sinistra

Vazokonstrikce



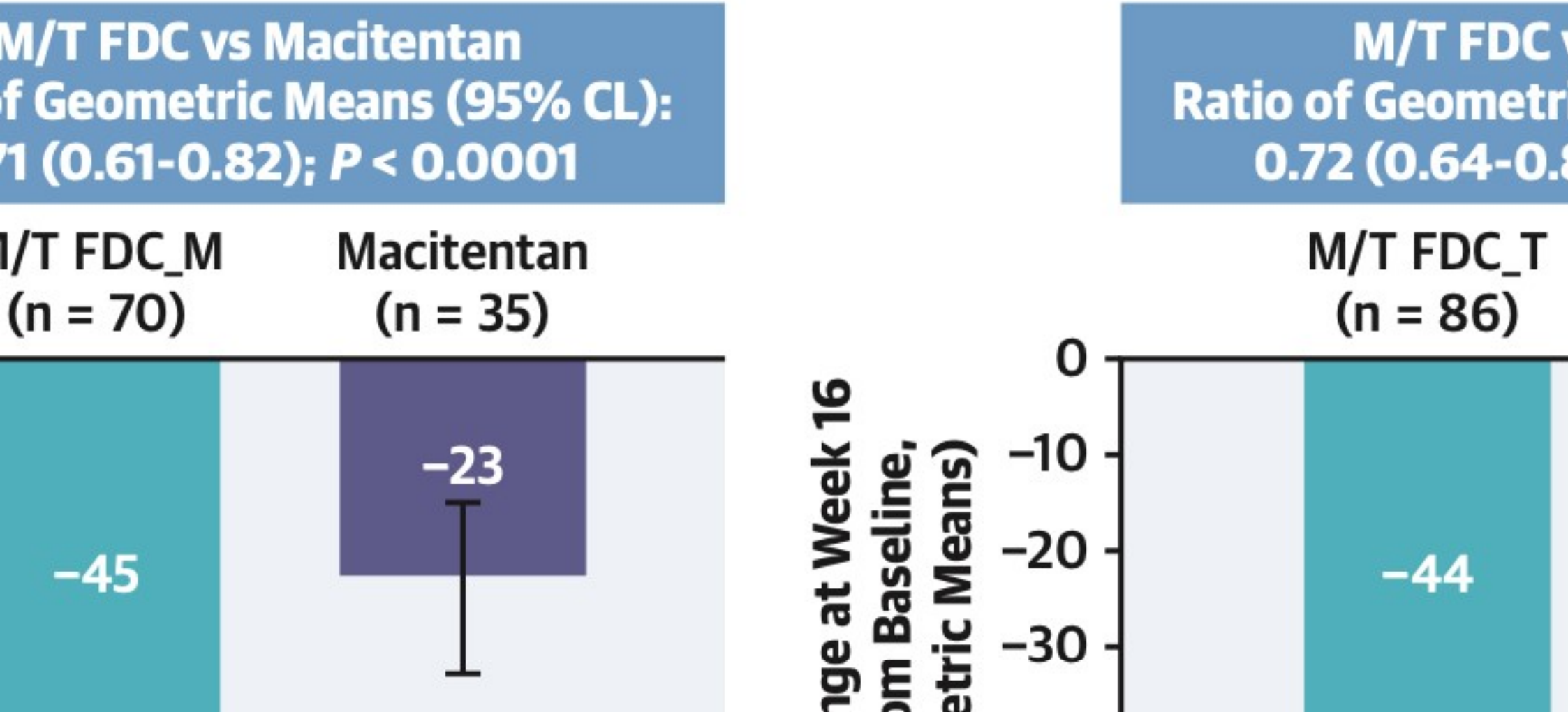


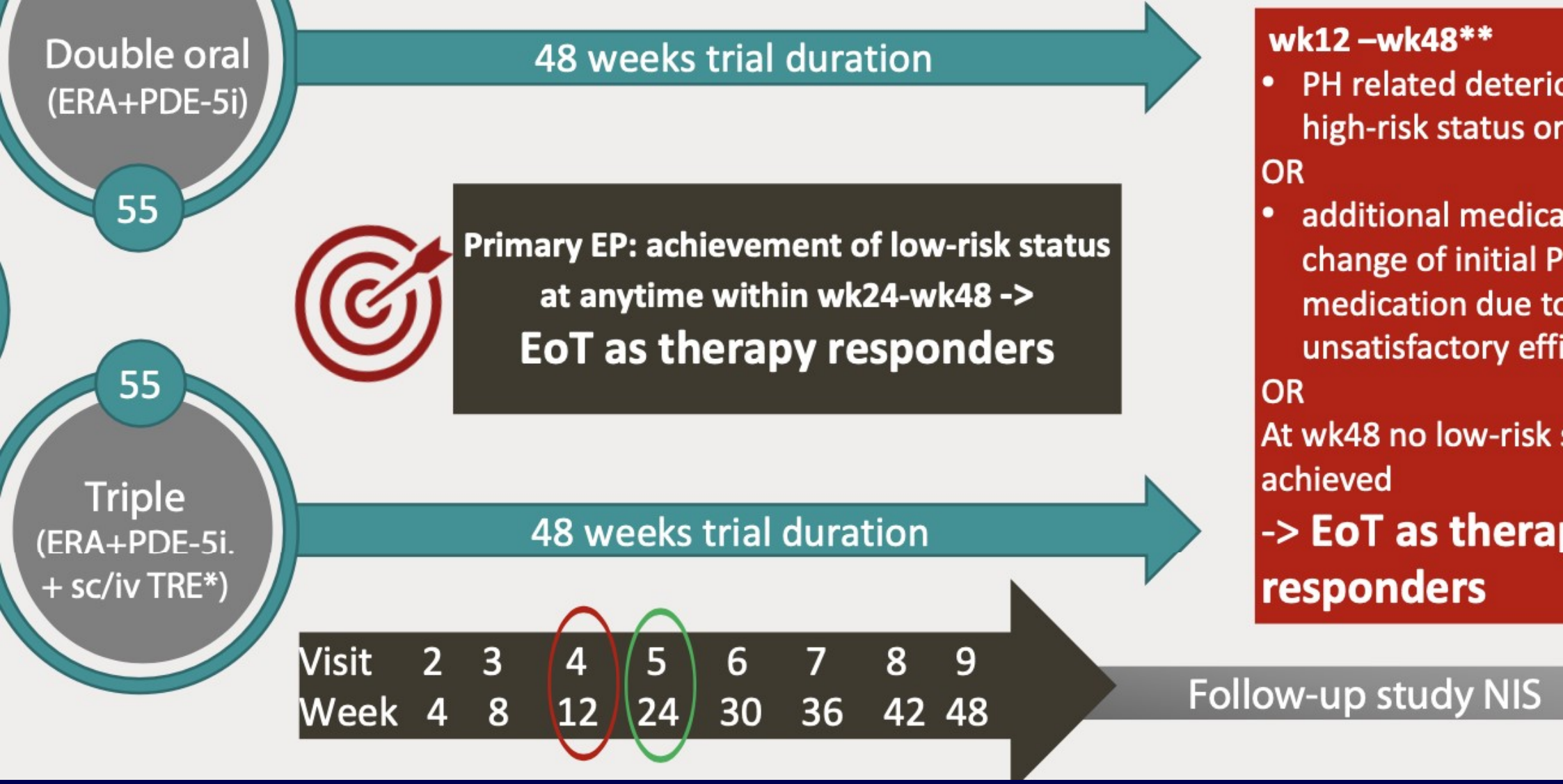




Grünig, MD,[a] Pavel Jansa, MD, PhD,[b] Fenling Fan, MD, PhD,[c] Jakob A. Hauser, MD, PhD,[d]
annaux, MSc,[e] Adele Morganti, MSc,[f] Hany Rofael, MD, PhD,[g] Kelly M. Chin, MD, MSCS[h]

monotherapy, single-tablet M/T FDC (n=108), macite



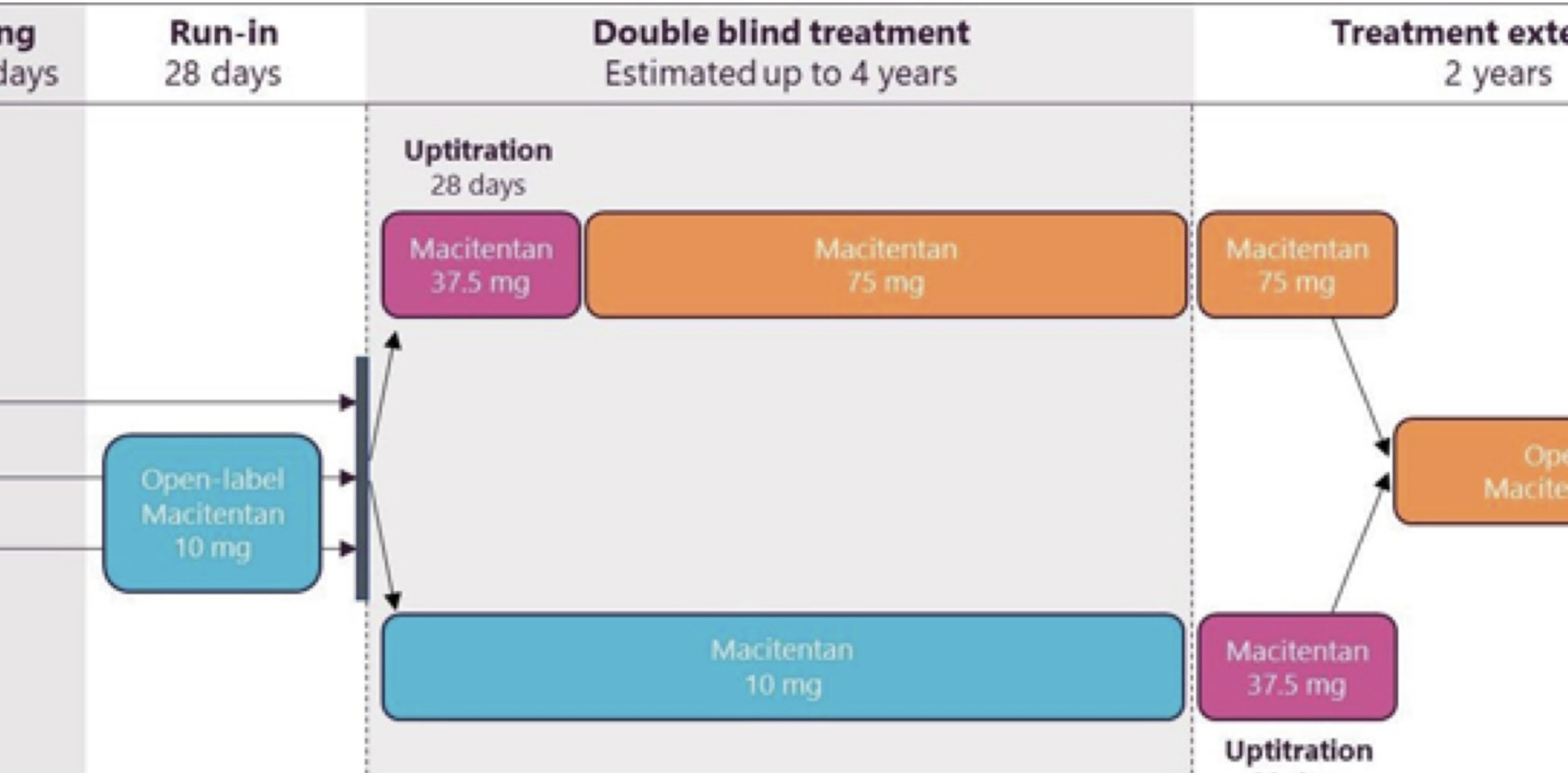


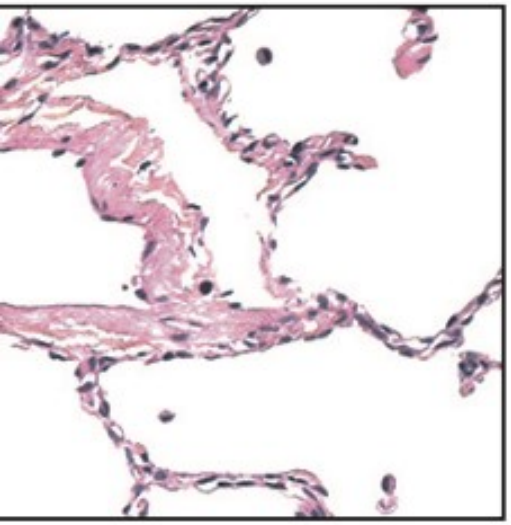
fety of initial triple therapy including parenteral treprosti
atients

ent

background PAH therapy, except for ERAs (must be s

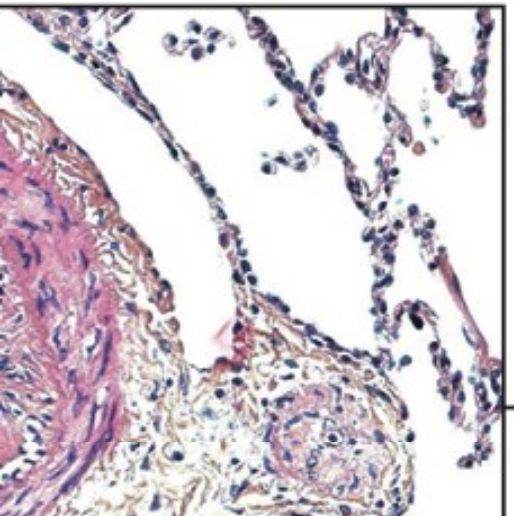
ead superiority study in PAH





Pulmonary vascular remodelling

ery



Dysfunctional pulmonary endothelium
- Imbalance between locally produced vasodilators and vasoconstrictors
- Excessive secretion of different factors influencing cell survival and growth
- Proinflammatory phenotype
- Changes in metabolic processes
- Acquisition of mesenchymal properties
- Decrease in tube formation

Dysfunctional vascular smooth muscle
- Sustained vasoconstriction
- Increased capacity to migrate, proliferate, and survive
- Changes in metabolic processes
- Altered expression, function, and regulation of ion channels
- Extracellular matrix remodelling
- Mobilisation and recruitment of progenitor cells

Persistent inflammation and immune dysregulation
- Overabundance of various inflammatory mediators, including of IL-1, IL-6, IL-8, IL-12,

Vasoconstriction:
↓ NO
↓ Prostacyclin
↑ ET-1

Deregulated TGF-β
↓ BMPR-II
↑ SMURF1
↓ pSmad1/5/8

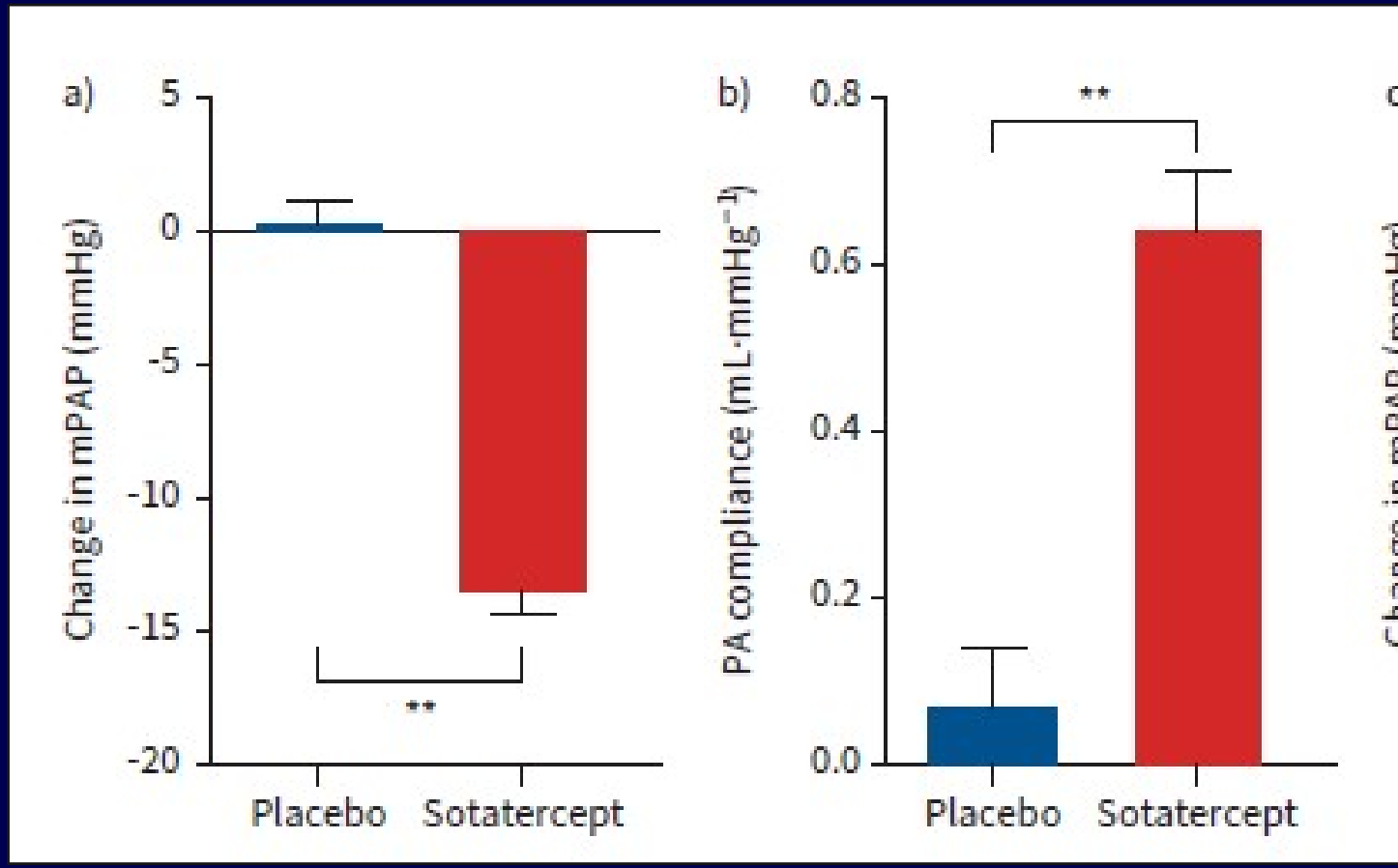
Growth mediators:
↑ FGF-2
↑ EGF
↑ PDGF

Extracellular matrix
↑ MMPs, TIMPs
↓ Elafin
↑ PAI-1

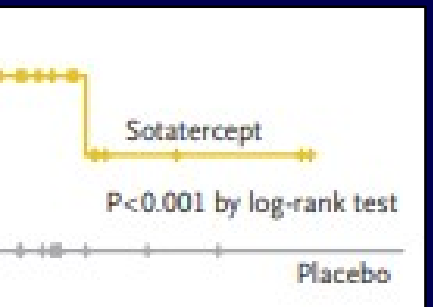
14.9 %, 24 weeks, NYHA FC II 49% + NYHA FC III 5

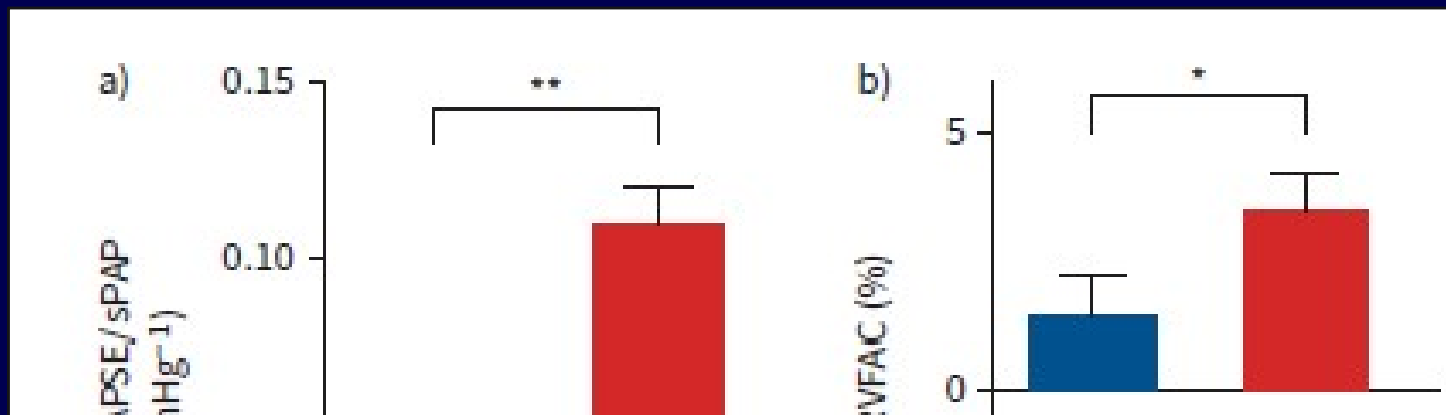
Hemodynamics

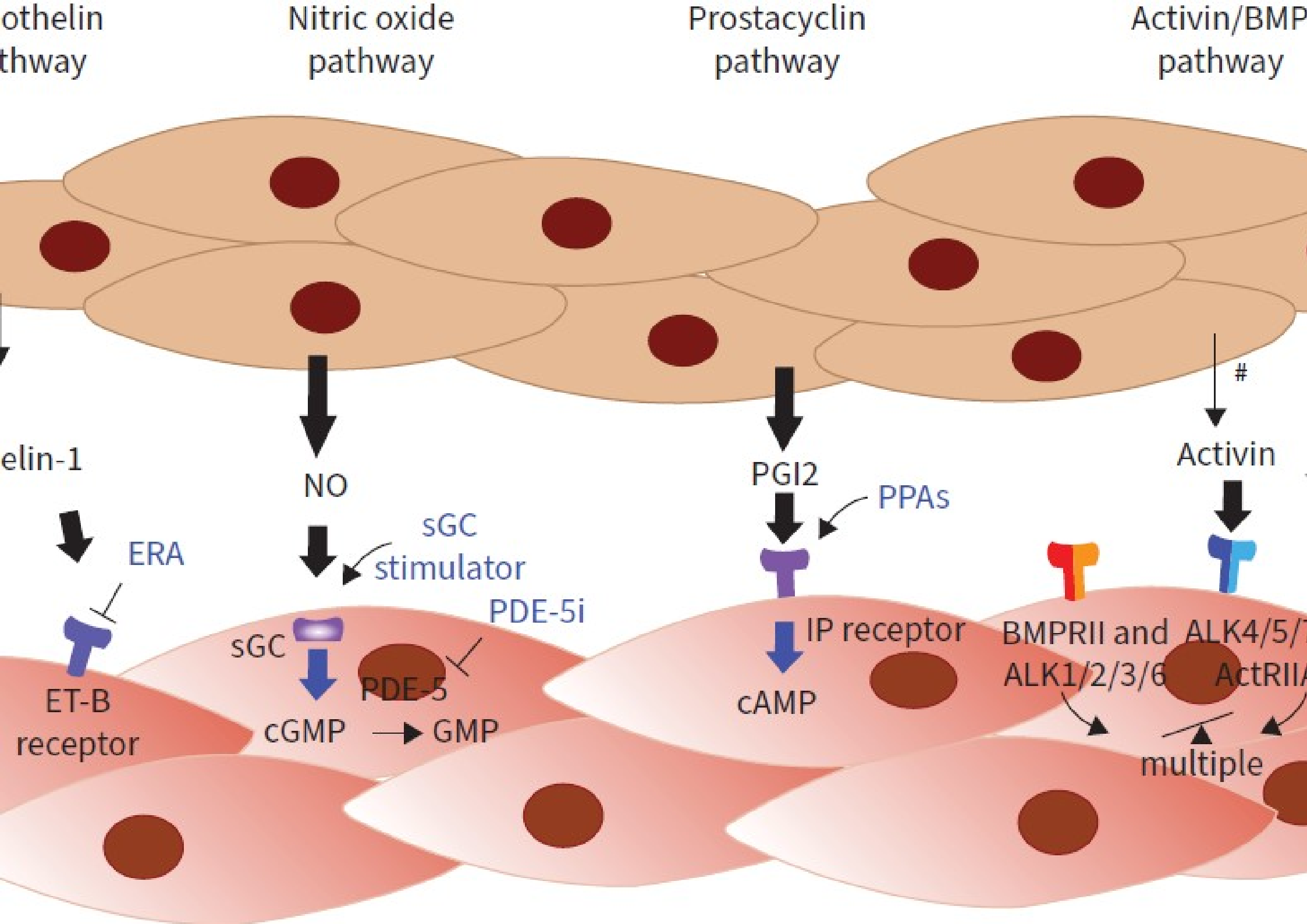
+40.1 vs. -1.4 m (p < 0.001)

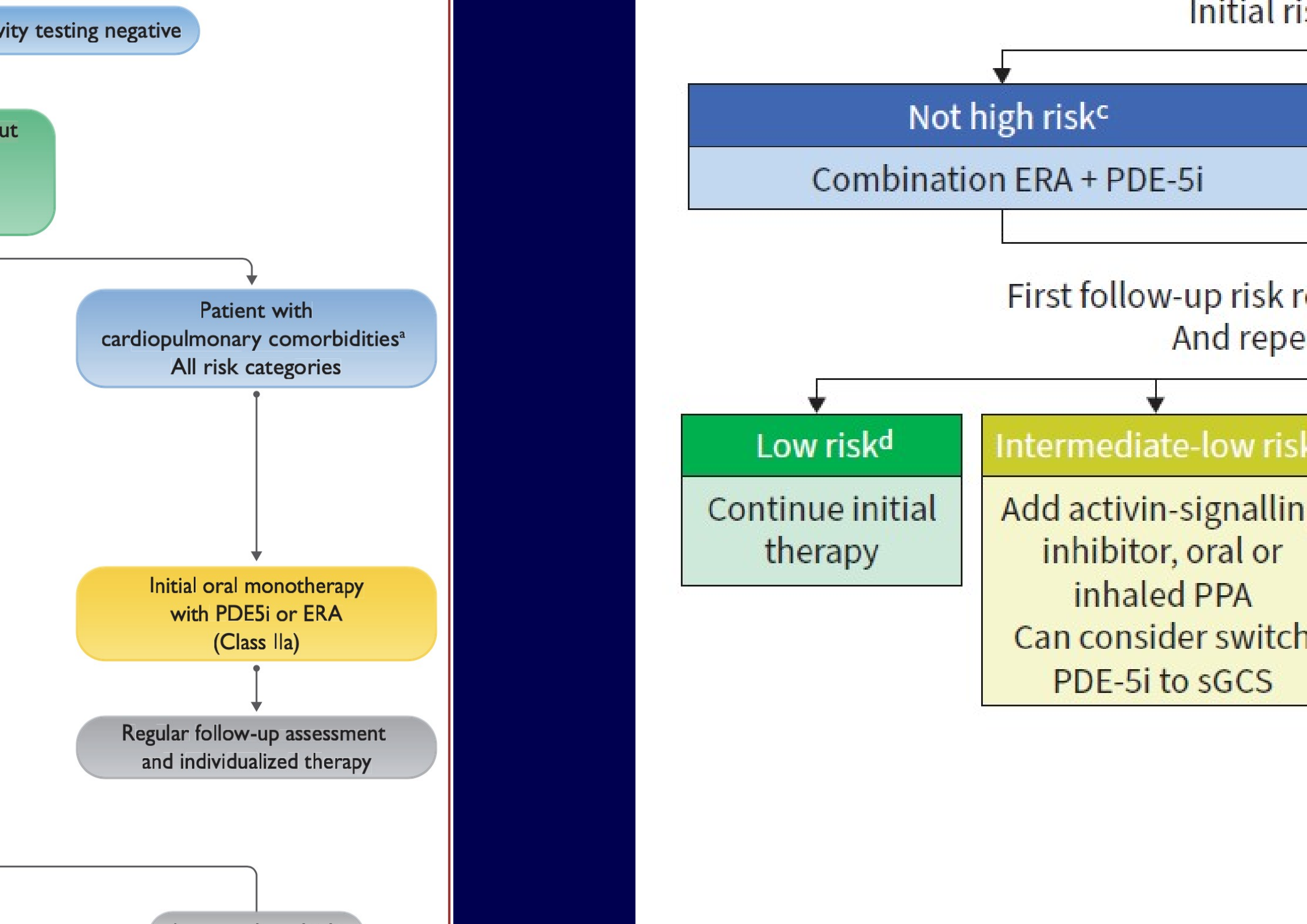


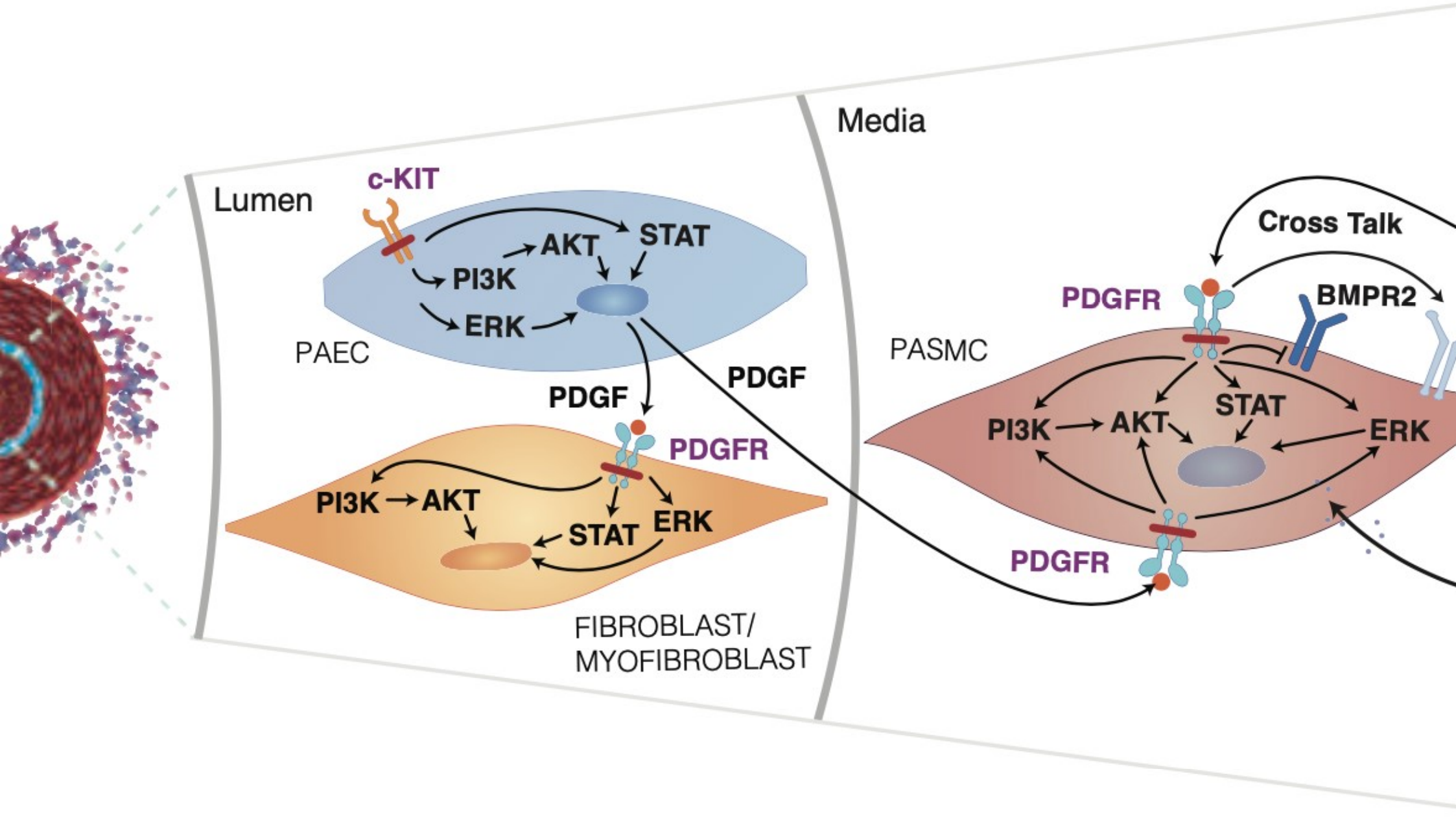
ECHO






othelin
thway
Nitric oxide
pathway
Prostacyclin
pathway
Activin/BMP
pathway
elin-1
ERA
ET-B
receptor
NO
sGC
stimulator
sGC
PDE-5i
PDE-5
cGMP
GMP
PGI2
PPAs
IP receptor
cAMP
#
Activin
BMPRII and
ALK1/2/3/6
ALK4/5/
ActRII
multiple


vity testing negative
ut
Patient with cardiopulmonary comorbidities[a] All risk categories
Initial oral monotherapy with PDE5i or ERA (Class IIa)
Regular follow-up assessment and individualized therapy
Initial ri
Not high risk[c]
Combination ERA + PDE-5i
First follow-up risk r
And repe
Low risk[d]
Continue initial therapy
Intermediate-low risk
Add activin-signallin inhibitor, oral or inhaled PPA
Can consider switch PDE-5i to sGCS

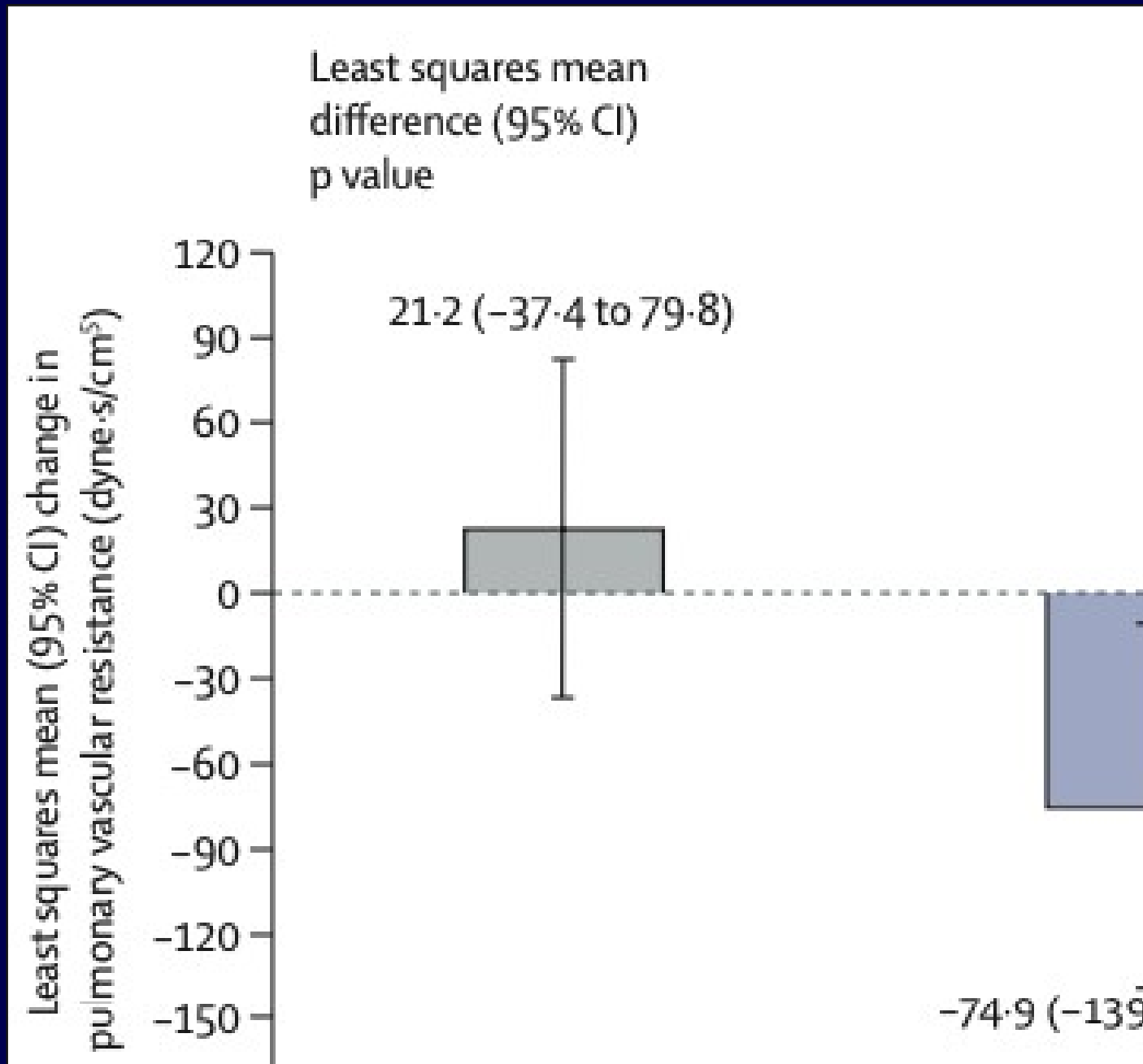

Lumen
c-KIT
AKT
STAT
PI3K
ERK
PAEC
PDGF
PDGF
PDGFR
PI3K
AKT
STAT
ERK
FIBROBLAST/
MYOFIBROBLAST
Media
Cross Talk
PDGFR
BMPR2
PASMC
PI3K
AKT
STAT
ERK
PDGFR

d to
nib

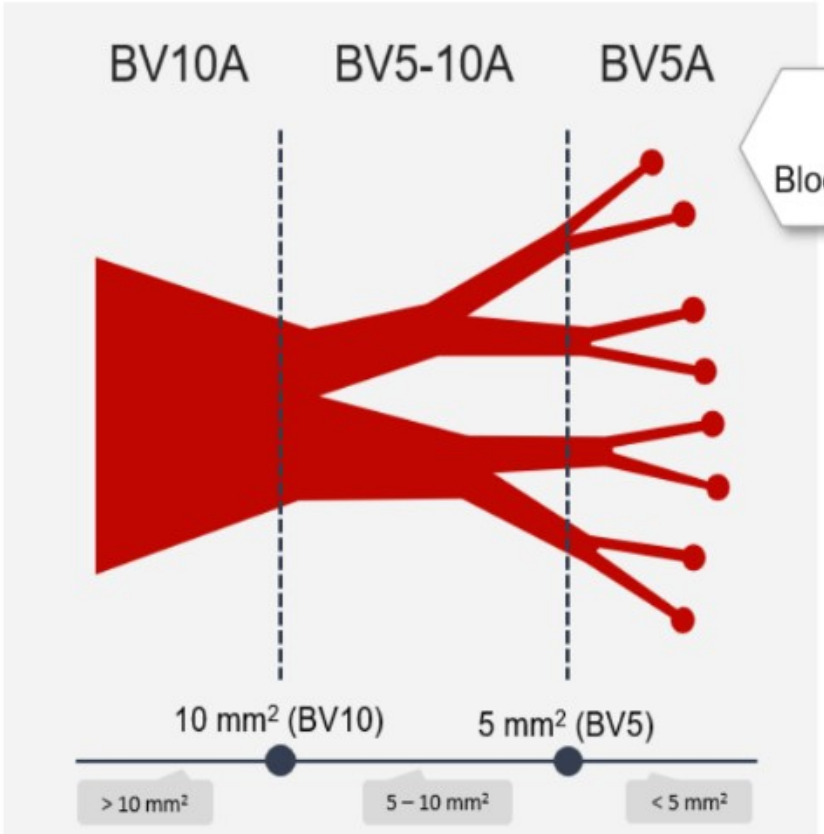
7 discontinued treatment
6 adverse events
1 protocol deviation

ed treatment

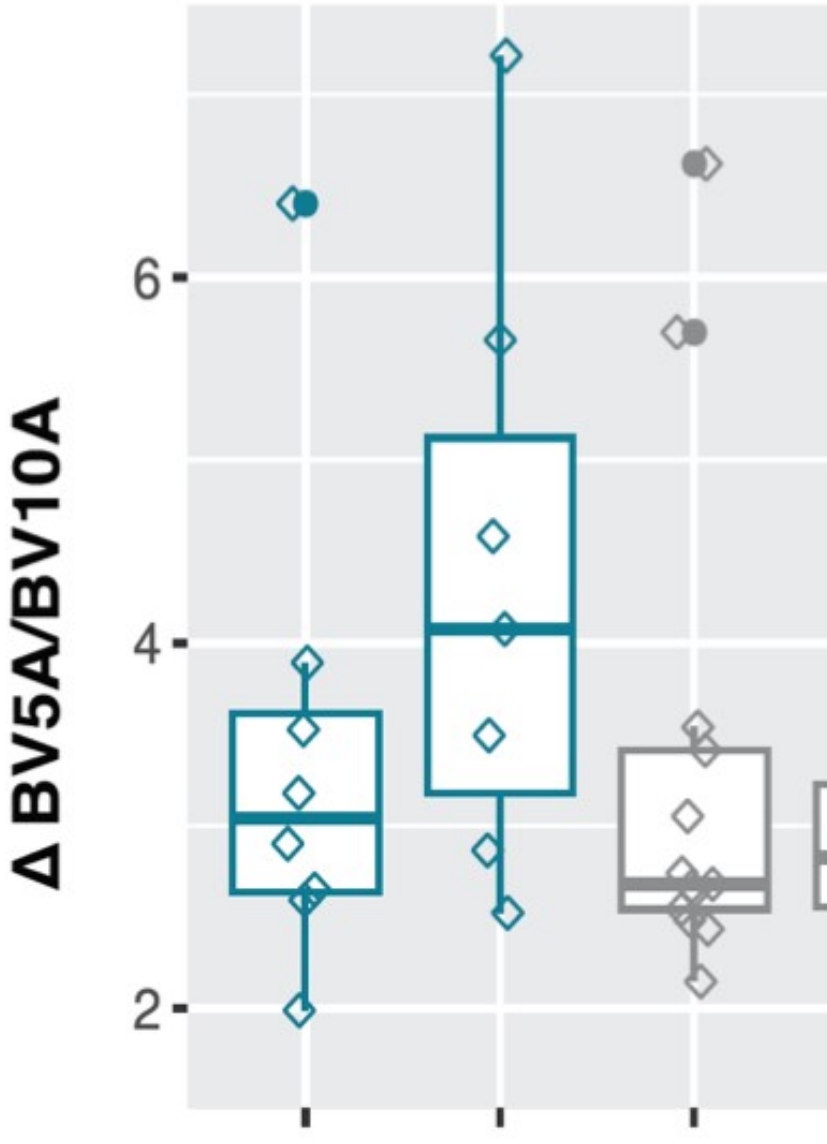
l in the

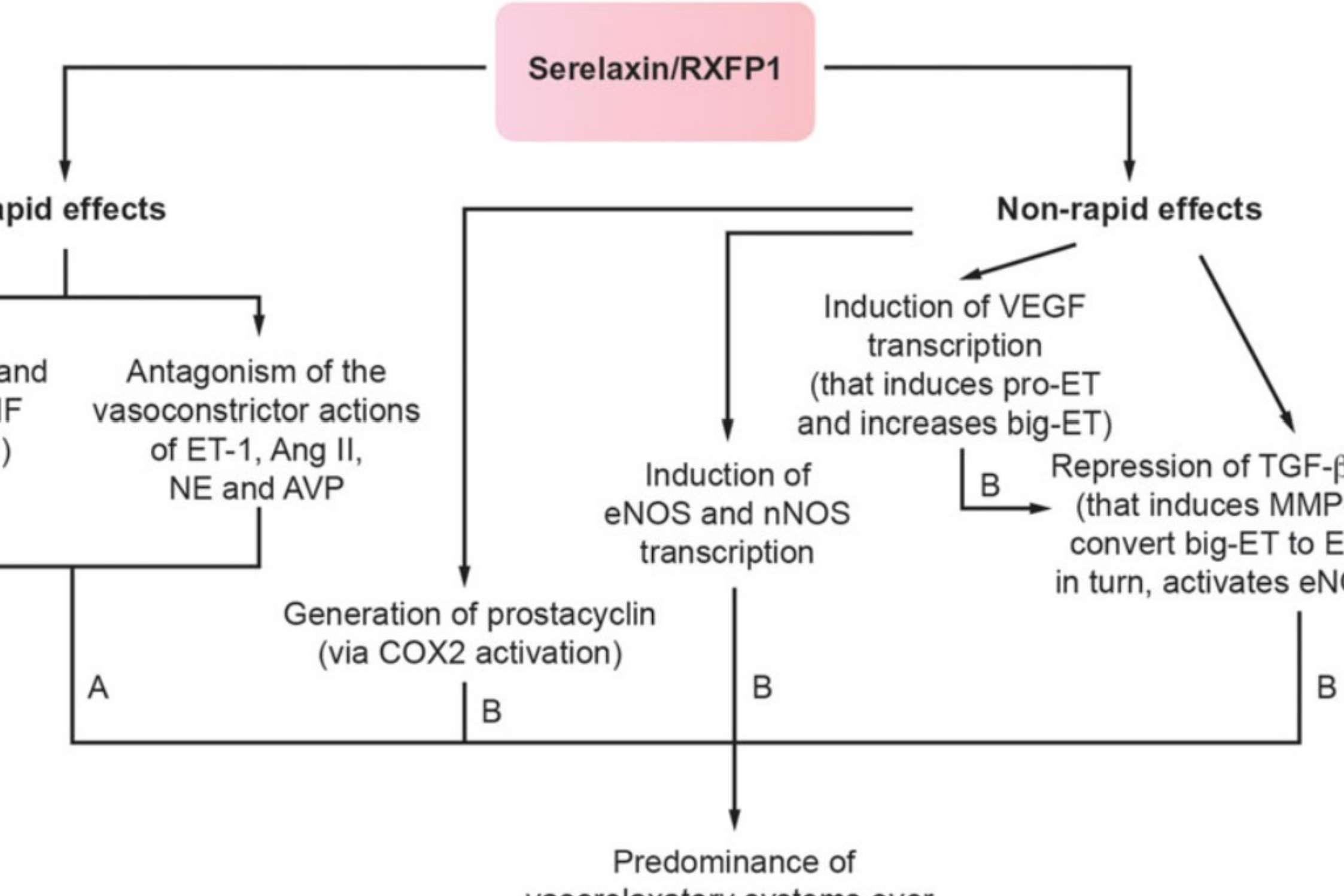


on and blood vessel volumes quantification



Parameter	L
BV5A/BV10A ratio	




Serelaxin/RXFP1
Non-rapid effects
Induction of VEGF transcription (that induces pro-ET and increases big-ET)
Induction of eNOS and nNOS transcription
Antagonism of the vasoconstrictor actions of ET-1, Ang II, NE and AVP
Generation of prostacyclin (via COX2 activation)
B
A
B
B
B
Predominance of

rticipants With Heart Failure and Pulmonary H

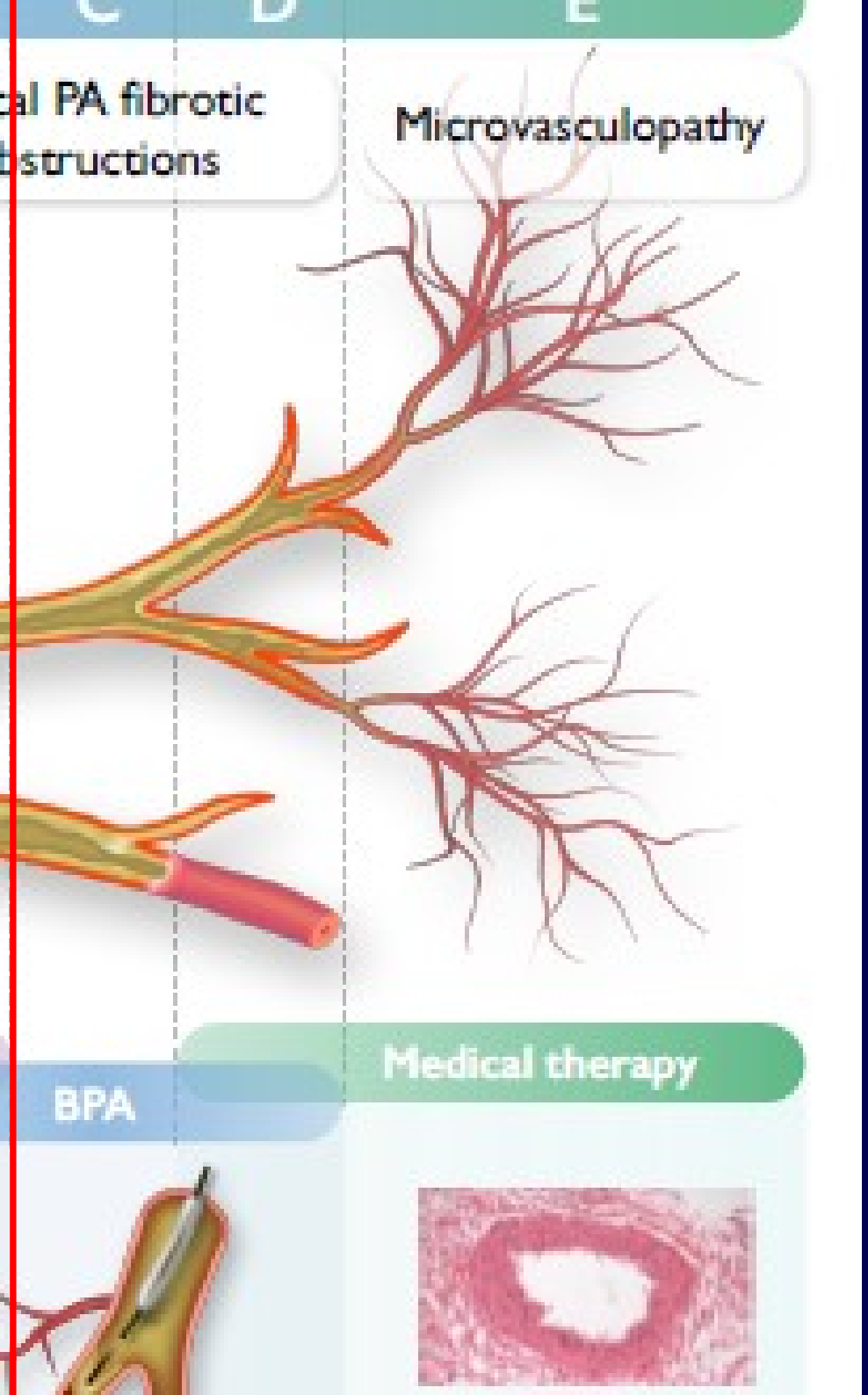
on date: 2025

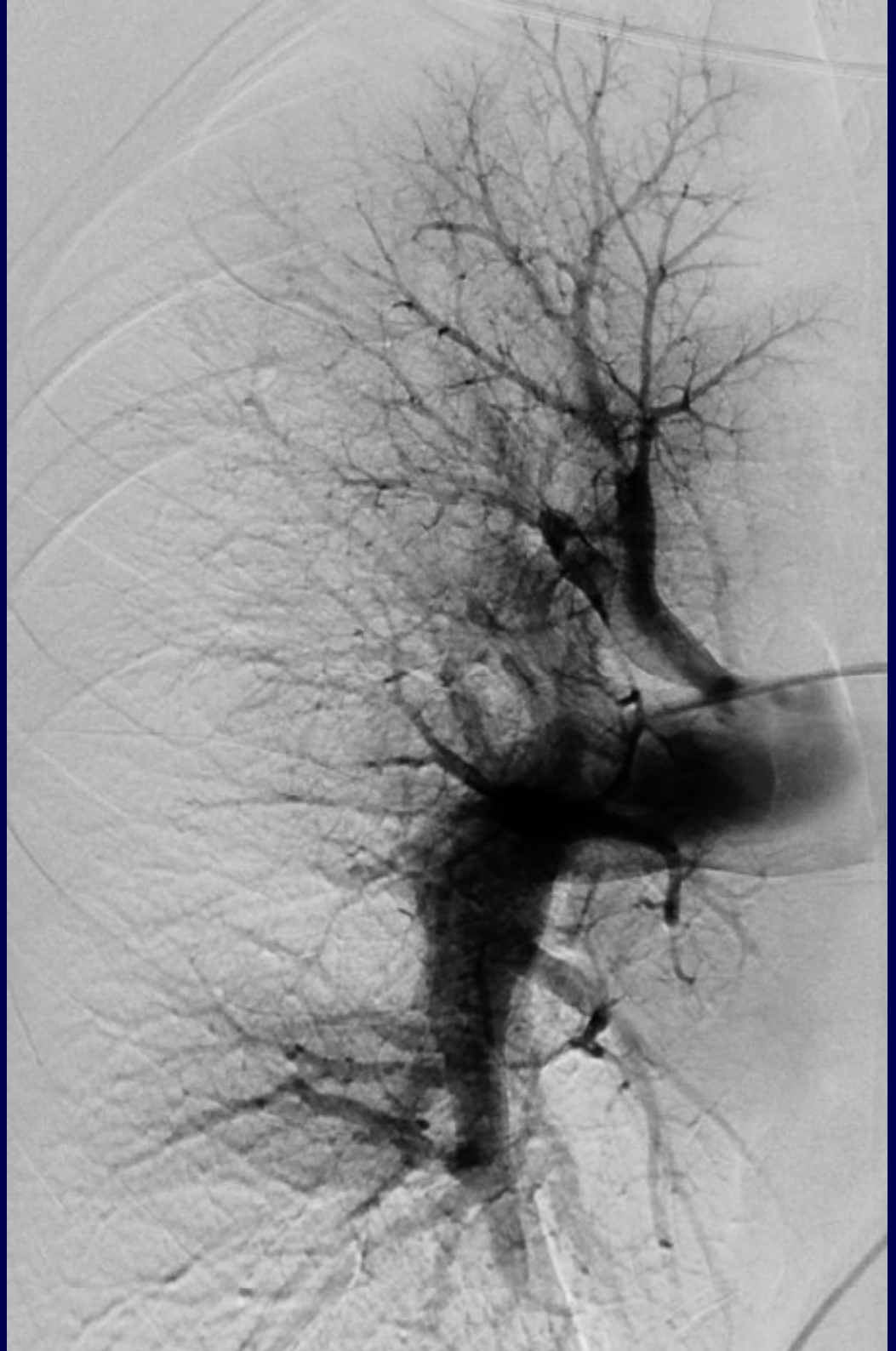
gle dose of either Dose A or Dose B or Dose C o

g diagnosis PH-LHD

(inhaled PDE5i in PH-IPF)
hib (inhaled PDGFR, CSF1R, c-KIT inhib
1 (inhaled imatinib in PH-ILD)
portable inhaled NO in PH-ILD)

A-PH COPD (inhaled sGC stimulator in
uat (inhaled sGC activator in PH-ILD)


C
D
E
al PA fibrotic
bstructions
Microvasculopathy
Medical therapy
BPA



A left A1, A8, A9, A10 a, b, c

A right A5, A8, A4, left A6a, b, A4, A5, A2

A right A10, A4, A5, A8, A9, A6, A, A2

	March 2020	Sep

n date: 2028

ticenter, prospective, randomized, controlled, op

H who are eligible for both PEA and BPA (randon

inary team konference, central adjudication com

bleted within 6 months from randomization.

at 4 months and 12 months after PEA or last BP

í terapeutické možnosti PAH vyvin
řípravků, 3 signální cesty) zlepšují
dní je ovlivnění pravé komory
evyléčitelné onemocnění
znivě ovlivňuje pozdní diagnóza a
zy:

VLADIMÍR DYTRYCH

Podpořeno firmou AOP

KOMPLEXNÍ
KARDIO
VASKULÁRNÍ

European
Reference
Network
for rare or low prevalenc
complex diseases

Network

vé vadě

bez jiných komorbidit, do té doby neužív

630m, B 0..0

20: 400 ng/l

ní stran PE

olicních žil, trikuspidální regurgitace 3-4+,

	PAMP (mmHg)	PCWP (mmHg)	CO (L/min)	
	39	7	3.83	

ekapilární PH – PAH asociovaná s vrozenou
í vadou s významným levo-pravým zkratem

FA pomalé dráhy 11/2020

notenství

nství a ponechání stávající léčby

nství a úpravu léčby

enství pokračovat

ris a **zahájena léčba subkutánním treprostini**
dávky 22 ng/kg/min ve 33. týdnu gravidity.

čba preventivní dávkou LMWH

yly po celou dobu gravidity v normě nebo jen leh

ýden gravidity): dobrá fce. LK, dilatace PK, kter
stacionární), odhad. PASP 64 mmHg, trikuspidá

anní porod přirozenou cestou

vaný porod císařským řezem

ogie VFN

nesteziologem naplánován **porod císařským ře**
aně 22.11.2021 v 37. týdnu gravidity.

kg/min a do léčby navrácen Volibris

ouhodobě v nízkém riziku při rizikové stratifikaci,
IP.

ila druhé těhotenství, ve kterém si přála pokračovat

ostupně navyšována dávka subkutánního treprostinilu,
35ng/kg/min

ušností NYHA II, s normálními nebo jen lehce zvýšenými
zn. pravostranného kardiálního selhávání

cí léčby, která byla eskalována v průběhu těh

skalace léčby

na až do současné doby kombinační léčba K

stinilem v dávce 85 ng/kg/min, která stejná jak

a ambulanci PH 8/2024:

75m, B 2..5

CTEPH po BPA	1
CTEPH nově diagnostikovaná v graviditě	1
PAH - vazorespondér	1
CTD-PAH	2
CHD-PAH	2
IPAH	5
PCH	1

ní těhotenství:

Porod živého plodu sekcí	14
Spontánní abort	1
Umělý abort	1

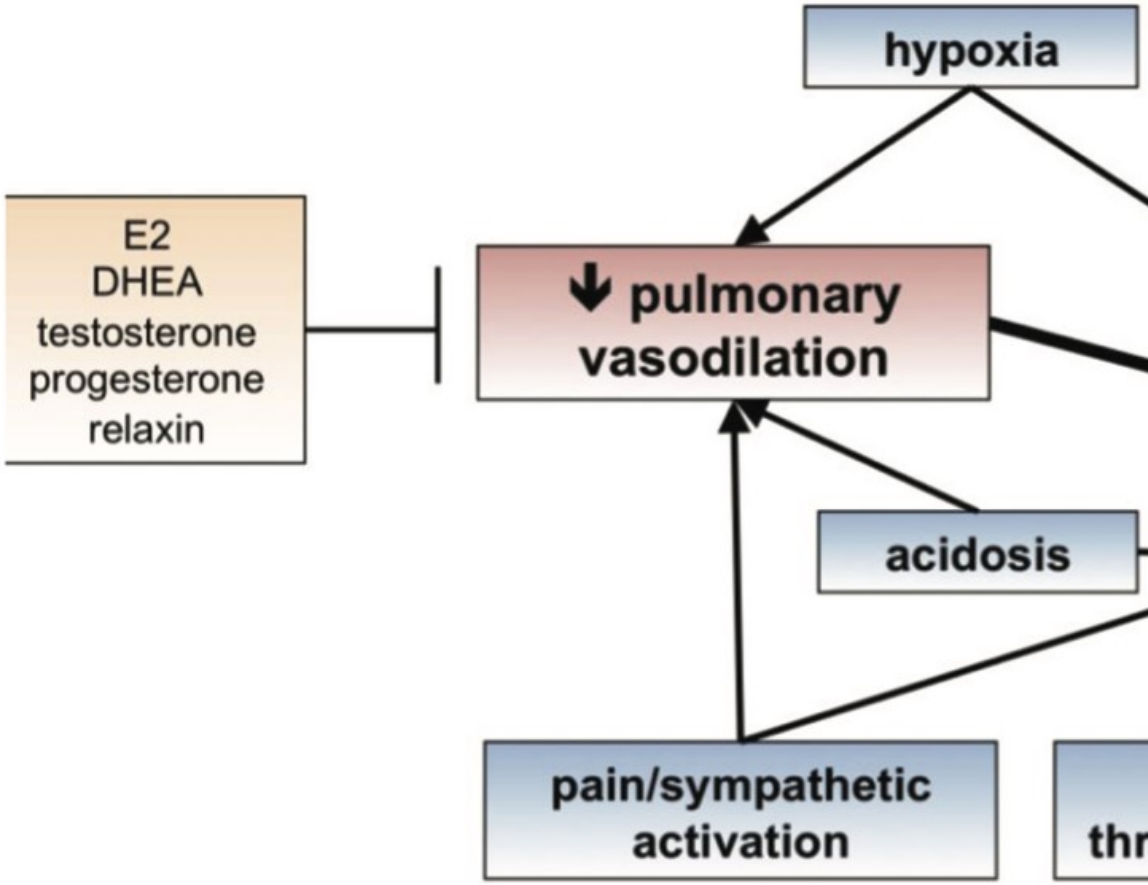
lní mortalita: 1 (33. d

– 50 %)

zistence až o 40 %

šti, u zdravých
P se nemění

nemůže přizpůsobit
P, PVR, afterload
elhávání PK během
po porodu



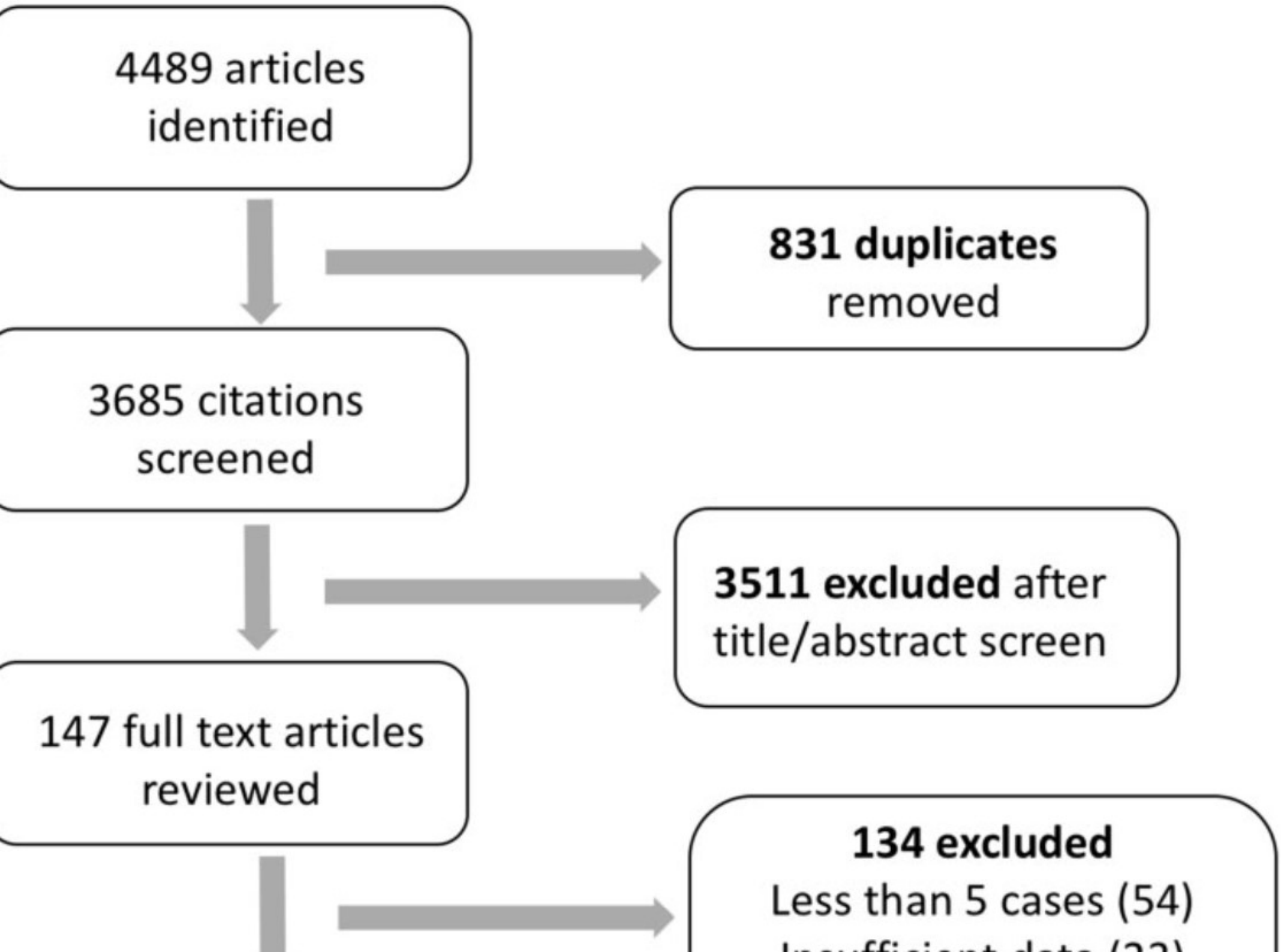
dělohou, role pohlavních hormonů

stav – vyšší riziko PE

kdy největší volumexpanze

é a tlakové změny (krevní ztráty vs. přesun extravas
ní utlačené VCI)

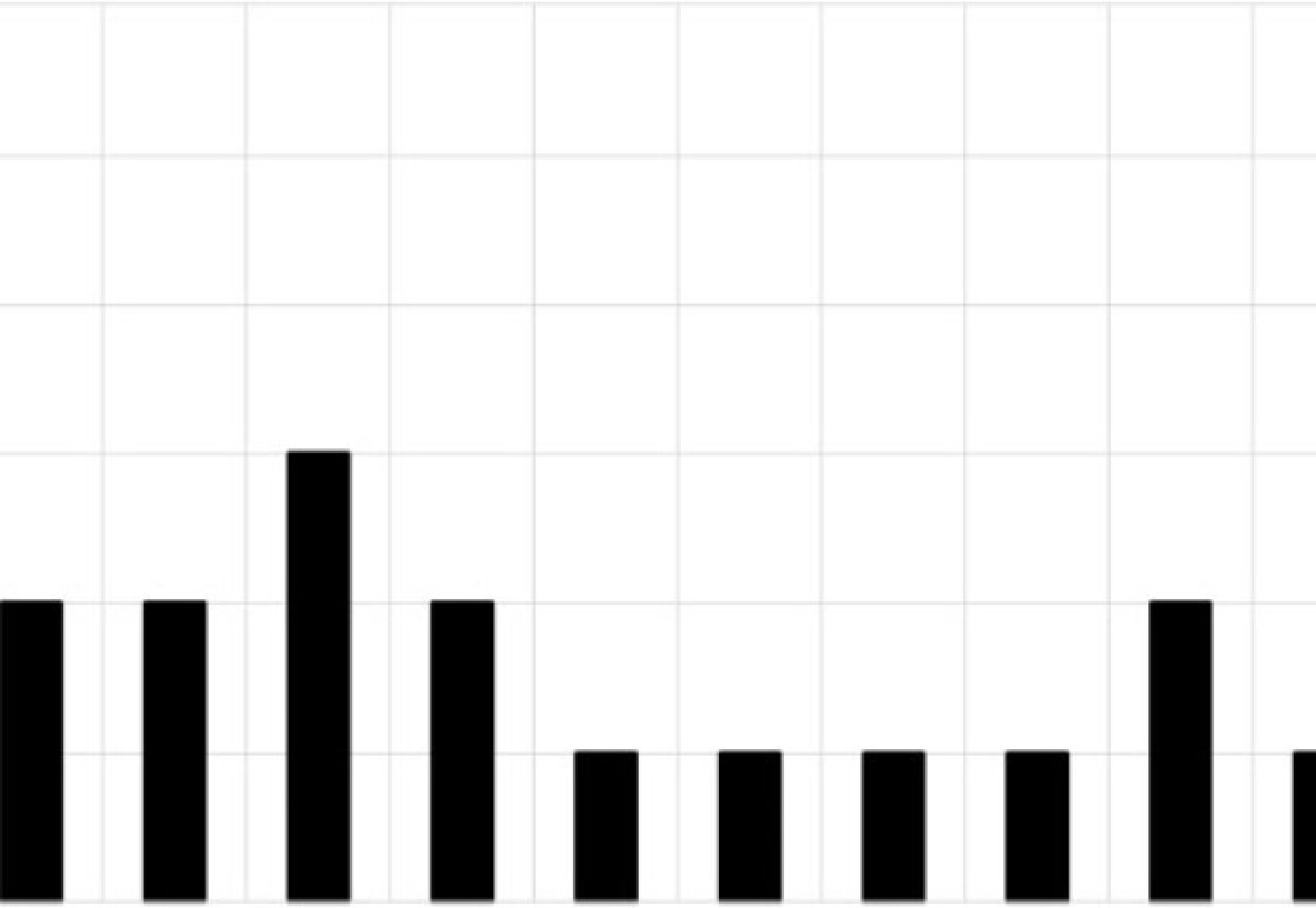
engerově syndromu – extrémní riziko, pokles sytém
vého zkratového proudění -> progrese hypoxie, puln
no selhání

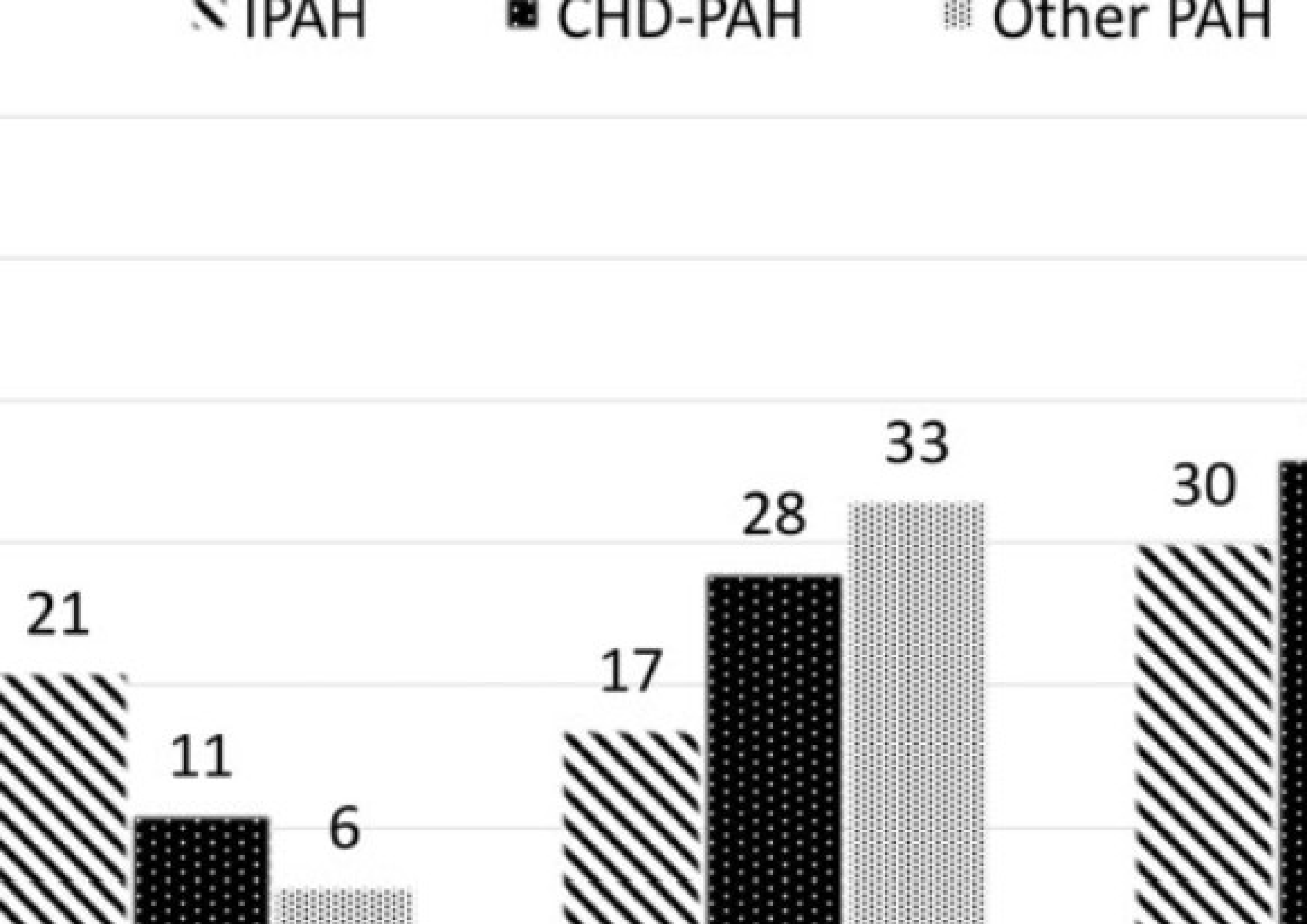

4489 articles identified
831 duplicates removed
3685 citations screened
3511 excluded after title/abstract screen
147 full text articles reviewed
134 excluded
Less than 5 cases (54)

Case collection	Country	Study type	Women (*n*)
2001–2015	USA	Retrospective	30
1997–2015	French	Retrospective	20
2010–2014	China	Retrospective	11
2008–2014	ROPAC	Retrospective	39
2007–2013	China	Retrospective	10
2006–2012	India	Retrospective	30
1999–2009	USA	Retrospective	18
–	USA	Retrospective	5
1982–2007	Japan	Retrospective	42
1995–2010	UK	Retrospective	7
2007–2010	US, Eu, Aus	Prospective	26
–	Israel	Retrospective	7

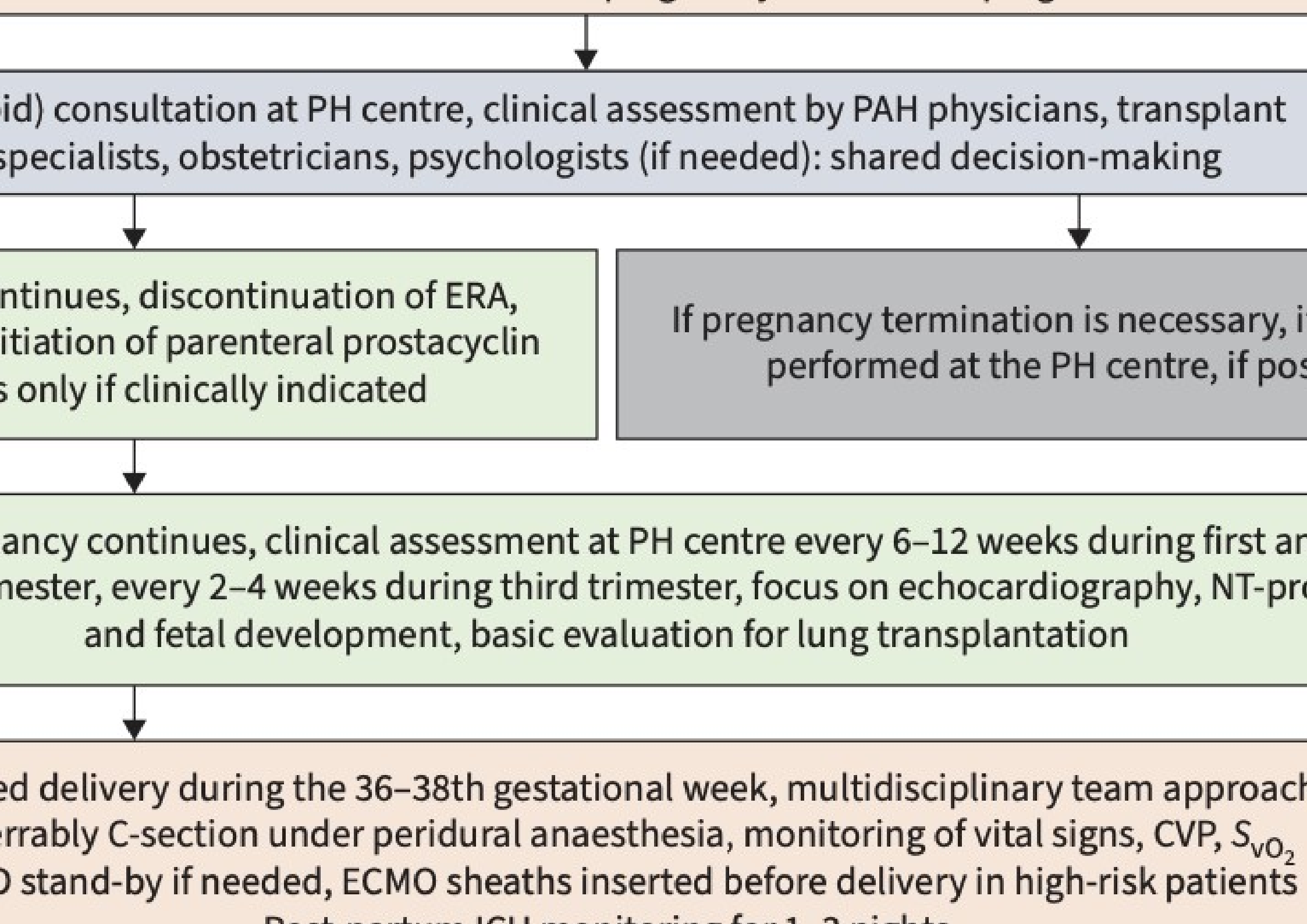
ics	Mean ± SD or %	Number of affected pregnancies	Total pregnancies in denominator
	28 ± 2	217	217
	60.3%	94	156
ure on echocardiogram (mmHg)	76 ± 19	154	154
	73.7%	126	171
	26.3%	45	171
	21.5%	46	214
	63.6%	136	214
	15.0 %	32	214
n)			
ancy	58.0%	79	136
	37.9%	25	66
	47.7%	92	193
	49.0%	51	104
	24.0%	25	104

nortality in PAH, *n* (%)	26 (12)
ity according to PAH etiologies	
AH, *n* (%)	9 (20)
eart disease-associated PAH, *n* (%)	15 (11)
iated PAH, *n* (%)	2 (6)
of death[a]	
or cardiogenic shock, *n* (%)	12 (6)
st or sudden cardiac death, *n* (%)	5 (2)
ypertension crisis, *n* (%)	2 (1)
a, *n* (%)	3 (1)
nfection/sepsis, *n* (%)	5 (2)
mbrane oxygenation, *n* (%)	6 (3)—4
ng transplant, *n* (%)	1 (0.5)—
fetal and neonatal outcomes	
nortality, *n* (%)	8 (4)
e, *n* (%)	6 (3)
ath rate, *n* (%)	2 (1)




IPAH
CHD-PAH
Other PAH
21
11
6
17
28
33
30

PAH (n=30), PH-LHD 1), PH-lung disease =1), CTEPH (n=7)	Delivery by C-section in 22/41 (54%) cases	8/49 (16%) post-partum deaths	5 al n
pregnancies): IPAH), CHD-PAH, (n=2), CTD-PAH (n=1)	C-section in 4/7 (57.1%) deliveries, vacuum-assisted vaginal delivery in the remaining cases	No peripartal maternal mortality (0/6)	N
(9 pregnancies), all PAH	C-section in 2/7 (28.6%) deliveries, 5 vaginal births	No peripartal maternal mortality (0/6)	T neo
PAH (n=12), PH-LHD , group 2 PH (n=45), 3 PH (n=4), group 5 PH (n=9)	C-section in 38/70 (58%) patients	No peripartal maternal mortality (0/70)	
pregnancies): IPAH 3), CTD-PAH (n=1), istosomiasis (n=1)	All deliveries by C-section (7/7)	No peripartal maternal mortality (0/5)	No
(17 pregnancies, 1 mini): IPAH (n=9), PAH (n=2), CHD-PAH (n=2)	All deliveries by C-section (17/17)	Maternal mortality 0/0 (0%), one patient required ECMO support and lung Tx	No (18/
(10 pregnancies): AH, (n=7), IPAH/HPAH	5/10 (50%) deliveries by C-section	Maternal mortality 0/0 (0%)	


id) consultation at PH centre, clinical assessment by PAH physicians, transplant
specialists, obstetricians, psychologists (if needed): shared decision-making
ntinues, discontinuation of ERA,
itiation of parenteral prostacyclin
s only if clinically indicated
If pregnancy termination is necessary, i
performed at the PH centre, if pos
ancy continues, clinical assessment at PH centre every 6–12 weeks during first an
nester, every 2–4 weeks during third trimester, focus on echocardiography, NT-pr
and fetal development, basic evaluation for lung transplantation
ed delivery during the 36–38th gestational week, multidisciplinary team approach
errably C-section under peridural anaesthesia, monitoring of vital signs, CVP, S_{vO_2}
O stand-by if needed, ECMO sheaths inserted before delivery in high-risk patients

torů hrají i např. osobnost pacientky, rodinné zázem

o být ponecháno na pacientce a partnerovi

u by měly být poučeny o nutnosti kontaktovat centru

čováno, pacientka by měla být intenzivněji sledová

léčba PAH zejména parenterálními prostanoidy